Neurosurgical considerations of cysticercosis of the central nervous system

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Infestation of the central nervous system (CNS) by the larval form of *Taenia solium* can be etiological for one or more of several clinicopathological manifestations. Experience gained from treating 18 patients forms the basis for a classification of this disease and for observations upon therapy. Twenty-three surgical procedures in 15 of the 18 patients provide the foundation for comment on operative treatment in management. Whereas most procedures are palliative, eradication of the CNS disease may be achieved in cases of solitary intraventricular cysts. There has been no operative mortality.

**KEY WORDS** • cysticercosis • *Taenia solium* • brain cyst • obstructive hydrocephalus • epilepsy • ventriculitis • arachnoiditis

Public health and sanitary engineering measures, governmental regulations, and usually high standards of personal hygiene in the United States assist in preventing infestation of man with the pork tapeworm (*Taenia solium*). Commonly known as cysticercosis, the larval stage of the cestode can develop in various tissues of the body after the oncospheres hatch from the eggs in the gut to penetrate the duodenum. The skin, muscles, heart, eyes and nervous tissue are common repositories in which, over months to years, the intermediate stage may develop clinical manifestations.

The migration of peoples from endemic areas to the United States results in the presentation of hitherto unfamiliar, or at least uncommon, clinical entities produced by the larval form of *T. solium*. Our experience gained from exposure to the neurological and neurosurgical challenges of this parasite is reported, together with a clinicopathological classification of this disease somewhat amplified beyond categories reported previously. For a general, but quite comprehensive, review of the disease, the reader is referred to Dixon and Lipscomb.9

**Clinical Material**

The case material upon which this communication is based was gathered in the UCLA Center for Health Sciences and the hospitals affiliated with the teaching program of the Division of Neurosurgery. The patients were treated between 1970 and 1980. Before 1975, sporadic cases of cysticercosis were operated on, but since 1975 the number of patients affected and the variety of central nervous system (CNS) involvement by the endoparasite have increased sufficiently to justify a study of this disease. All of the 18 patients in our experience came to the United States from either Mexico or South America.

The clinicopathological manifestations are complex and do not lend themselves easily to simple classification, a not unexpected feature in view of the protean nature of the larval behavior in man. While recognizing such limitations, a system of classification is presented which accommodates each of the patients we have treated and is in general conformity with reports published previously. Previous classification systems are amplified by the addition of ventriculitis as Category IV, a hitherto unemphasized form of the disease. This tabulation omits involvement of the peripheral nervous system, of which there appears to be only one report.16 The value of this attempt to classify CNS cysticercosis may rest solely on its succinctness in displaying the range of clinicopathological behavior of the etiological agent (Table 1).

The case material of this series is distributed into
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TABLE 1
Classification of central nervous system cysticercosis

<table>
<thead>
<tr>
<th>Disease Category</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>diffuse parenchymatous disease without focal mass</td>
</tr>
<tr>
<td>I</td>
<td>disseminated larval death inflammatory reaction</td>
</tr>
<tr>
<td>II</td>
<td>toxic encephalopathy &amp; meningitis</td>
</tr>
<tr>
<td>III</td>
<td>calcified larval form with seizures</td>
</tr>
<tr>
<td>IV</td>
<td>basilar adhesive &amp; racemose form</td>
</tr>
<tr>
<td>IV</td>
<td>obliterator arachnoiditis/meningitis hydrocephalus mixed types with cisternal cysts; spinal disease</td>
</tr>
<tr>
<td>V</td>
<td>intraparenchymatous cysts as mass lesions solitary as part of multifocal disease</td>
</tr>
<tr>
<td>VI</td>
<td>subarachnoid &amp; cisternal cysts local symptomatology mixed types with ventriculitis, intraventricular, &amp; intraparenchymatous cysts</td>
</tr>
<tr>
<td>VII</td>
<td>intraventricular cysts solitary multiple mixed types with ventriculitis &amp; cisternal cysts</td>
</tr>
<tr>
<td>VIII</td>
<td>spinal forms arachnoiditis/meningitis extra- &amp; intramedullary forms</td>
</tr>
</tbody>
</table>

TABLE 2
Incidence of disease by categories in 18 patients in this series

<table>
<thead>
<tr>
<th>Disease Category*</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>0</td>
</tr>
<tr>
<td>II</td>
<td>4 (2 also in Cat. V)</td>
</tr>
<tr>
<td>III</td>
<td>1 (also in Cats. VI &amp; VIII)</td>
</tr>
<tr>
<td>IV</td>
<td>3 (1 also in Cat. VI, I in Cat. VII)</td>
</tr>
<tr>
<td>V</td>
<td>3 (2 also in Cat. II, 1 in Cat. VI)</td>
</tr>
<tr>
<td>VI</td>
<td>4 (1 also in Cats. II &amp; VIII, 1 in Cat. IV, 1 in Cat. V)</td>
</tr>
<tr>
<td>VII</td>
<td>9 (also in Cat. IV, 1 surgically unconfirmed)</td>
</tr>
<tr>
<td>VIII</td>
<td>1 (also in Cats. II &amp; VI)</td>
</tr>
</tbody>
</table>

* For classification see Table 1.

TABLE 3
Operative procedures performed for cysticercosis on 15 patients, 1970 to 1980*

| Procedure                                                      | No. of Cases |
|                                                               |             |
| ventricular shunting                                          | 6           |
| suboccipital craniotomy with cyst evacuation                  | 10          |
| frontal transventricular cyst evacuation                      | 1           |
| suboccipital explorations (ventriculitis only)                | 2           |
| temporal craniotomy for incisural cyst evacuation             | 1           |
| frontal or frontotemporal craniotomy for intraparenchymatous cyst evacuation | 2 |
| parietal craniotomy for intraparenchymatous cyst evacuation   | 1           |

* Twenty-three procedures were performed.

Experience with Each Category of Cysticercosis

Diffuse Parenchymatous Disease

There were no certain examples of Category I (diffuse parenchymatous disease) among the 18 patients in this series. Therefore, one cannot comment on it other than to suggest that steroid therapy may improve the management of the inflammation that is presumed to follow larval death in cases of extensive infestation. This thought is based upon our experience utilizing successful, symptomatic, steroid treatment in one patient with extensive disease of Categories IV and VII plus recurrent encephalopathy. Current methods of monitoring and treating increased intracranial pressure may offer an advance beyond measures heretofore described, including surgical decompression.9,19,26

Calcified Larval Form with Seizures

Two of our four patients in Category II (calcified larval form with seizures) also had intraparenchymatous larval forms acting as mass lesions, a not unexpected association. Although epileptic attacks represent one of the commonest presenting symptoms associated with this affliction,1 the management of these cases is rarely surgical unless the epilepsy is symptomatic of a space-occupying process. In our cases without mass lesions, anticonvulsant therapy was the mainstay. Such patients have a very high incidence of intracranial calcification as demonstrated by plain radiological study; however, the greater diagnostic capabilities of computerized tomography (CT) scanning in identifying the usually multiple sites of larval deposits are confirmed by our experience.

Basilar Adhesive and Racemose Form

Fortunately, we have had to care for only one patient in Category III (basilar adhesive and racemose form). The pathological process defies surgical cure, but, in concert with others, we employed cerebrospinal fluid (CSF) diversion to manage the hydrocephalus.
Surgical experience gained from exploring this one patient (who also harbored multiple subarachnoid-space and cisternal cystic lesions) supports the generally dismal prospects of permanent help. The operative note reads: "The cysts blended into multiple arachnoidal adhesions which ensnared the optic nerves, carotid artery and posterior cerebral artery. . . . The entire clivus region was encased in a milky white arachnoid network. . . . Several areas were filled with multiple small cysts which washed out during the dissection." Unhappily, this kind of chronic basilar adhesive meningitis may accompany otherwise more promising manifestations of the disease, such as intraventricular cysts, and render an initially hopeful outcome quite otherwise. Such have been the findings of others with a wider experience.\(^3,5,12,14,15,17,21,25\)

**Ventriculitis**

Not previously identified as a variety worthy of separate distinction, Category IV (ventriculitis) is set apart because of our experience from two of our three patients who met the criteria. The first of these was a 37-year-old woman of Mexican birth whom we had treated over the preceding 2 years for hydrocephalus associated with a cystercerosis meningitis (mild CSF pleocytosis and positive titers to the parasite). She had also had multiple cystic masses removed from the cerebellopontine angle. She was temporarily aided in each instance by her shunting procedure and suboccipital craniotomy, but she returned with a 1-week history of nausea, vomiting, vertigo, and nystagmus. Although known to have a right Sylvian cyst, she was found to have a large fourth ventricle, highly suggestive of an isolated and enlarging ventricle or an intraventricular cyst. At exploration, the midline foramen of the aqueduct was shown to be blocked from above and (in contrast with filling of the fourth ventricle prior to her shunting procedure) the ventricle did not fill with oxygen from below and was mildly dilated on CT scanning. At suboccipital exploration, a mild arachnoiditis of the great cistern was noted, with partial obstruction of the midline foramen by adhesions. There was a greenish-yellow thickening of the ventricular lining compatible with ventriculitis. The aqueduct was occluded and no CSF entered the ventricle. Her symptoms improved, but her superior aqueductal syndrome persisted during the 6 months of most recent follow-up examination.

The second of the three patients was a 24-year-old man, who had presented with headache, nausea, vomiting, papilledema, and a stiff neck. A working diagnosis of cystercerosis was made, and a ventriculoperitoneal shunt procedure was performed, followed by prompt improvement. Within 6 weeks her headache returned, and she developed a superior aqueductal syndrome (Parinaud's syndrome). The aqueduct was shown to be blocked from above and (in

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**Intraparenchymatous Cysts as Mass Lesions**

Of our three patients in Category V (intraparenchymatous cysts presenting as mass lesions), two also had calcified lesions (Category II), and one suffered from subarachnoid and cisternal cysts as well. There is evidence from the literature that such lesions may be solitary.\(^9,14,18\) so far as the nervous system is concerned, but such solitary disease is quite rare.\(^12,17,20,26,27\)

In the absence of other telltale radiological signs in the brain (the patient reported on by Dixon and Lipscomb\(^6\) and subsequently by Ray\(^18\) had calcified lesions in her extremity musculature), intraparenchymatous mass lesions simulate tumors of other etiologies. In each of our three patients, the intraparenchymatous cystic lesion appeared as an enhancing ring-like tumor on CT studies, and there was no angiographic evidence of precise etiology (Fig. 1). If clinical indications exist for excision, such as in the case of a solitary lesion, or, in multifocal disease, if the lesion is symptomatic, then total excision is recommended (Fig. 2). This recommendation is based on the observation of remnants of the wall of the larval form of T. solium that were removed at operation at this institution and were successfully cultured. Small pieces (without the scolex) re-form a bladder, grow in length and width, and can be seen to undergo slow undulations and contractions.\(^6\) We have found, however, the spillage of the cyst contents has not had
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deleterious consequences. Within the brain substance, the cyst may be quite adherent as a result of an inflammatory reaction, as commented upon by Dyck, et al. 

**Subarachnoid and Cisternal Cysts**

Three of the four patients with lesions in Category VI (subarachnoid and cisternal cysts) had manifestations of one or more other categories. The one patient without other indications of disease was a 56-year-old woman from Ecuador with a 6-month history of ataxia and mild dementia. She was demonstrated to have severe hydrocephalus and a cystic lesion of the right cerebellar hemisphere. At craniotomy a cyst, at least 2.5 cm in diameter, was found in the subarachnoid space above the right hemisphere of the cerebellum, extending to the incisional notch. It was delivered, unruptured, by gentle teasing dissection. The patient made an excellent recovery. The other three patients were operated on for symptomatology related to the locale of other cystic lesions. Thus, cysts were removed from the prepontine (clival) region, the cerebellopontine angle, the lateral medullary recess, and the retro-pulvinar area. Symptomatic improvement could be documented, but other evidence of disease precluded any thought of cure.

Most of these subarachnoid cysts occur beneath or at the tentorial margin, and they often are multiple. Surgically, they are delivered by gentle traction and hydraulic dissection. They are usually unimpeded by any firm attachments. They tend to conform to the surgical egress route and at times almost “flow” into the exposure. When they are multiloculated or enmeshed in arachnoiditis, their delivery can be more tedious. Use of the Valsalva maneuver by the anesthesiologist may assist in delivering lesions in deep recesses and in revealing multiple lesions not previously suspected.

**Intraventricular Cysts**

The largest group, nine of our patients, had intraventricular cysts (Category VII). Although some of

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FIG. 1. Computerized tomography scan after contrast administration of an intraparenchymatous larval form of *T. solium*. Note the low-attenuation pattern with a halo of edema and a thin ring of enhancement defining the cyst. A characteristic calcified sign of a dead form is noted in the opposite hemisphere.

FIG. 2. Histological preparation of larval form of *T. solium* excised from the right frontal lobe of a 9-year-old girl with seizures, headache, nausea, and a small ring-like enhancing mass. The undulating “wall” is characteristic of the parasite, and the scolex could be identified on higher magnification. H & E, × 28.
these patients had multiple cysts, and some also fell into other categories of the disease; sufficient numbers had solitary cysts so that we entertained hopes of curing the neural aspects of such patients. Seven of our nine patients harbored presumably solitary CNS cysts (one at the interventricular foramen of Monro and six in the fourth ventricle). The eighth patient of the group suffered multiple cystic disease of the ventricle plus ventriculitis. The last patient in this category, a 24-year-old Mexican, is an unconfirmed case. He suffered progressive bifrontal headache, lethargy, and signs of involvement of the superior aqueduct with loss of upward gaze and nystagmus retractorius. Lesions were demonstrated within the third and fourth ventricles, with hydrocephalus due to the fourth ventricular obstruction. A ventriculoperitoneal shunt, only, was performed.

The villous exterior surface of the cyst wall, the presence of muscle bands on the interior surface, and the undulating movements noted on culture suggest a migratory life style of the cyst. If, as is presumed, the oncospheres gain entrance into the ventricular cavities via the choroid plexuses, cysts should be encountered in any ventricle. This has been our observation. In patients with multiple cysts, surgical procedures are likely to be but palliative. If the lesion is solitary, the motility of the lesion may explain the predominance of a fourth ventricular location. These concepts of downward migration gain support from the analysis of radiological studies by Dorfsman, the surgical findings of Hernandez, the postmortem studies of Sato, and our own experience.

Unlike the examples of highly complex management problems presented by the mixed and disseminated forms of the disease, the solitary, intraventricular cases are relatively straightforward. The single patient with a lesion in the foramen of Monro presented with unilateral ventricular dilatation after a 1-month history of suboccipital headache and vertigo with postural change. He showed severe papilledema and fine horizontal nystagmus. The lesion was removed as one would a colloid cyst in the same area. It was delivered into the lateral ventricle by vigorous irrigation, and the patient made an excellent recovery.

The other six patients with "solitary" cysts possessed similar medical histories. All had headache, which was mainly occipital, often severe, and at times intermittent and at times influenced by postural changes. Vomiting was common. The symptoms occurred over a 4-week to 11-month time span. Papilledema, nystagmus, ataxia, and dysarthria occurred sporadically, but objective neurological abnormalities could be meager. Peripheral blood eosinophilia was found twice but not given heavy weight in the diagnostic formulation. The radiological findings were quite uniform. Hydrocephalus existed, with an enlarged fourth ventricle on CT scanning. The fourth ventricle was usually, but not always, disproportion-
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FIG. 4. Computerized tomography scans of the posterior fossa. A: Scan with intravenously infused contrast medium reveals no enhanced outline of the enlarged fourth ventricle and its contents. B: After positive-contrast ventriculography, the thin rim of contrast material clearly identifies the low-attenuation boundaries of the cystic mass filling the ventricle.

ally large compared with the rest of the ventricular system. Positive-contrast ventriculography combined with CT provided reasonably secure evidence of the presence of only a single cyst in the four cases where it was used. The definition of a cyst as the cause of the enlarged ventricle was offered either by the use of positive-contrast ventriculography utilizing conventional radiography, or by CT scanning, or both (Fig. 3). The intraventricular cysts, in contradistinction to those located within the parenchyma of the brain, appeared only as low-attenuation areas without boundary definition except that of surrounding normal tissue. No enhancement was demonstrated with contrast administration (Fig. 4).

The operative approach was by way of a midline suboccipital exposure. The meninges were normal in all cases, and some tonsillar herniation was documented. The cyst could present at the midline foramen, but division of the inferior vermis was required in some instances. The mass could present as a glistening, golden-green or whitish, smooth-walled cyst, and on occasion would be delivered almost spontaneously as the tonsils were separated (Fig. 5). Hydraulic dissection, aided by gentle Valsalva maneuvers, often sufficed. Flat-bladed forceps helped to ease the lesion out of the ventricle. In most instances, the lesions were evacuated without rupture. Rupture, however, has not been associated with adverse reactions, meningitis, or scolex dissemination, such as may occur with an Echinococcus cyst. In this regard, it was suggested to the author by Dr. Gonzales-Cornejo that our immunity from reactions secondary to cyst rupture could be explained by our quite regular use of intraoperative steroid medication. Dr. Aguirre-Por-tillio also advised the author that he has encountered

FIG. 5. Suboccipital craniectomy with delivery of a solitary fourth ventricular larval form of T. solium via an enlarged midline ventricular opening.
a severe inflammatory reaction following rupture of a fourth ventricular cyst, and that the reaction responded very favorably to steroid administration. As in the case of intraparenchymatous cysts, and for the reasons noted, all fragments of any broken cyst should be removed. All of our patients with a solitary fourth ventricle cyst recovered well.

Our experience with solitary cysts in a clinic in the United States (albeit through patients of non-native origin) is in accord with reports from other geographic areas, as well as with a smaller series in the United States reported by Latovitzki, et al. Thus, the surveys from Romania, South America, Mexico, Spain, and Poland indicate that solitary cysts are encountered in only 8% to 25% of a symptomatic population.2,3,14,17,20 The fourth ventricle is the usual site for such solitary lesions, and this form of the disease yields the best results.

Spinal Forms

Only one of our patients could qualify for Category VIII (spinal form of infestation). At the same time this patient suffered from disseminated disease of which the spinal manifestations were prominent although not amenable to surgical treatment. Cystic lesions, hard masses on the surface of the spinal cord, and intramedullary lesions occur, the latter being the most common. These are approachable by operation. A recapitulation of the world literature concerning this form is to be found in the recent report by Akiguchi, et al.1

Diagnostic Considerations

The protean behavior of the infestation prevents a simplistic generalization with respect to diagnosis. The site or sites of the larval forms determine the symptoms and the findings, whether these be hydrocephalus, seizures, local mass effects, or inflammatory reactions.

The radiological approach, often the initial investigation, yields information that may raise the index of suspicion as to the etiology of a given illness. The radiological findings have been well discussed by prior authors.4,7,8,10,23 In our hands, the analysis of peripheral blood or CSF has rarely been comprehensive, but an eosinophilia in either should attract the clinician's attention. (We were reminded that eosinophils in the CSF will not be identified unless a Wright's stain is utilized.) We have been disappointed in the value of serological testing: the delay in obtaining results blunts the application for early diagnosis. The indirect hemagglutination test (routinely used at the Center for Disease Control) and the indirect immunofluorescent tests are the best.22 There is no doubt, however, that an even better serological test is needed.

Of greatest value to us has been knowledge of the patient's country of origin, even after a lapse of several years. In our area, a Spanish surname offers a clue to the suspecting physician, and it should be a clue to others when seeing patients from areas in which this disease is endemic.

Treatment

There is as yet no satisfactory therapeutic treatment to eradicate all vestiges of the infestation. The surgical approach is one of treating symptomatic manifestations and, in a minority of patients (those with solitary cysts), this may possibly be curative. Each patient deserves study for adult worm infestation as part of the clinical assessment so he may be purged of dangers of auto-reinfestation, and so measures can be taken to protect the public health. As of the present time, the sanitary engineering and public health approach will have the widest influence in eliminating the disease.

References


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J. Neurosurg. / Volume 55 / September, 1981
Cysticercosis of the central nervous system


__Manuscript received February 11, 1981.__

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