Surgical considerations in treatment of intraventricular cysticercosis

An analysis of 45 cases

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Infestations of the human brain with the larval stage of Taenia solium, once an infrequent diagnosis in the United States, is now a more frequently encountered clinical entity especially in population centers with high immigrant flux. During a recent 5-year period 45 cases of intraventricular cysticercosis have been evaluated and treated. Modes of involvement included isolated cyst formation, ependymitis, or combinations of both. Evidence of associated parenchymatous involvement was present in 20% of cases. Sites of infestation included the lateral ventricle (five cases), third ventricle (12 cases), Sylvian aqueduct (four cases), and fourth ventricle (24 cases). Mean post-therapy follow-up periods for this series exceed 36 months. This experience indicates that direct excision is the treatment of choice for ventricular cystic lesions, but that management, operative planning, and expectations should be influenced by considerations of: 1) the potential for acute clinical deterioration (38%); 2) the potential for cyst migration; 3) attendant ependymitis, defined by computerized tomography or verified at surgery; 4) the potential for increase in cyst volume with local mass effect; 5) selection and institution of corridors of surgical access that establish alternative routes of cerebrospinal fluid flow; and 6) the possibility of cyst excision by a stereotaxic endoscopic procedure.

Key Words • intraventricular tumor • brain cyst • cysticercosis • obstructive hydrocephalus • computerized tomography • stereotaxic surgery

Cerebral cysticercosis was initially observed in man by Panarolus in 1650, in a priest who died following a convulsive episode.1,18 That individual manifested cystic lesions of the corpus callosum. It was not appreciated for many years that a parasitic form had caused such cyst formation. In 1855, Küchenmeister31 fed cysticerci to a condemned murderer and noted specimens of Taenia solium in the individual’s intestine at autopsy, thus providing the first evidence that Cysticercus cellulosae was indeed the larval form of the pork tapeworm. Many pathological and clinical reports have followed, offering further definition to our concepts related to the pathophysiology and management of this complex disorder.2,5-35,37-56 The majority of these communications have emanated from countries where the disease is endemic, such as Chile, Poland, Spain, Mexico, Africa, and Asia.

Neurocysticercosis was described in the United States by Walter Dandy in 1927.15 Since then, exposure and reports of the disease have been infrequent until recently when immigrant activity from areas endemic for the disorder has increased.50,53 Concurrently, recent technical advances in imaging and surgical technique have caused reappraisal of diagnostic and therapeutic concepts related to this infestation.5,21,39,47,57

Although the larval stage of the cestode may become localized within various neurocompartments, including the brain parenchyma, subarachnoid space, spinal cord, and ventricular system, its occurrence within the ventricular system may be particularly hazardous, creating a substrate for acute excursions in intracraniatal pressure (ICP).18,42,53 To date, strict definition for management of the spectrum of intraventricular infestation in light of the current armamentarium of imaging and surgical capabilities has not been realized.

This report details our recent 5-year experience in the management of 45 patients with primarily isolated intraventricular involvement.

Clinical Material

The case material used as a basis for this report was gathered from the neurosurgical services at the Los Angeles County-University of Southern California Medical Center during a 5-year period terminating in
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TABLE 1

*Summary of 45 cases of intraventricular cysticercosis*

<table>
<thead>
<tr>
<th>Location of Lesion</th>
<th>Type of Lesion</th>
<th>Initial Treatment</th>
<th>Secondary Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>lateral ventricle (5)</td>
<td>cystic (5)</td>
<td>transcallosal excision (4)</td>
<td>none</td>
</tr>
<tr>
<td>third ventricle (12)</td>
<td>cystic (9)</td>
<td>stereotaxic (1)</td>
<td>none</td>
</tr>
<tr>
<td>aqueduct of Sylvius (4)</td>
<td>cystic with ependymitis (1)</td>
<td>ventriculostomy (2)</td>
<td>none: died</td>
</tr>
<tr>
<td>fourth ventricle (24)</td>
<td>ependymitis (2)</td>
<td>transcallosal interformenial excision (6)</td>
<td>none</td>
</tr>
<tr>
<td></td>
<td>stenosis (ependymitis) (4)</td>
<td>transcortical excision (1)</td>
<td>none</td>
</tr>
<tr>
<td></td>
<td>cystic (17)</td>
<td>transcortical (1)</td>
<td>VP shunt (1)</td>
</tr>
<tr>
<td></td>
<td>cystic with ependymitis (7)</td>
<td>VP shunt (2)</td>
<td>none</td>
</tr>
<tr>
<td></td>
<td></td>
<td>VP shunt (4)</td>
<td>none</td>
</tr>
<tr>
<td></td>
<td></td>
<td>excision (11)</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td></td>
<td>VP shunt (6)</td>
<td>cyst excision (2)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>excision (6)</td>
<td>VP shunt (6)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>VP shunt (1)</td>
<td>cyst excision (1)</td>
</tr>
</tbody>
</table>

* Numbers in parentheses indicate number of cases. VP = ventriculoperitoneal.

October, 1982. All of the 45 patients in this series were Mexican or Central American in origin. There were 24 females and 21 males, with an age range of 10 to 62 years (mean 34 years). All individuals underwent computerized tomography (CT) assessment of their intracranial cavity as well as positive-contrast ventriculography. Immunoassays were obtained uniformly, with diagnosis being established on histological assessment or convincing patterns of epidemiological, immunological, and radiographic findings considered indicative of such an infestation. All cases were considered eligible for classification under Disease Category IV or VII as defined by Stern; however, none demonstrated evidence of associated cisternal or arachnoid involvement. Minimal parenchymatous disease was observed in 20% of our cases.

**Lateral Ventricular Cysts**

Five patients who presented with complaints of chronic headache, nausea, and vomiting were found to have isolated cystic lesions within the lateral ventricular region (Table 1). Two of these were admitted in coma without focal signs following rapidly progressive courses. No parenchymal or temporal horn involvement was observed in any of these patients. Three cysts were clearly shown to be mobile on contrast ventriculography. One 2.5-cm third ventricular cyst had migrated to the right lateral ventricle during pneumoencephalography.

All five lateral ventricular lesions were successfully and uneventfully excised, four via a transcallosal approach and the fifth by CT-guided stereotactic endoscopy.* The latter procedure employed a Brown-Roberts-Wells stereotaxic system and was accomplished using a 6.8-mm endoscopy system with continuous irrigation of the region with Ringer's lactate solution. The patient had been treated with high-potency glucocorticoids. No further operative therapy has been required in any of these patients during periods of observation lasting a mean of 30 months.

**Third Ventricular Cysts**

Twelve patients had cysts in the third ventricle (Table 1). Although 10 patients were symptomatic, with periods of headache lasting from 2 weeks to 8 months, two individuals had abrupt courses with acute headache progressing to lethargy within hours and demanding prompt action. Two other patients died suddenly while being evaluated on medical services for chronic headache, in spite of rapidly placed bilateral ventriculostomies. Both were found to have simple cystic lesions measuring 2.0 and 2.5 cm in the third ventricle, without evidence of ependymitis or more complex disease.

All of the remaining 10 patients demonstrated at least minimal evidence of ventriculomegaly and underwent ventriculostomy for monitoring and control of ICP; bilateral ventriculostomies were performed in the event that bilateral foramen of Monro obstruction was evident on contrast ventriculography. Two patients manifested radiographic evidence of posterior third ventricular ependymitis and veins, with occlusion of the outlet at the iter. These patients were treated successfully with ventriculoperitoneal (VP) shunts.

Direct surgical exploration was undertaken on eight patients with cystic lesions; a transcallosal approach was effected in six patients and a transcortical route in two. In all cases a generous fenestration of the septum pellucidum was accomplished to assure adequate bilateral ventricular drainage via a unilateral approach. One patient manifested ependymitis at the foramen of Monro and anterior compartment of the third ventricle. This patient had previously shown contrast enhancement on CT scanning in this region. Ventriculoperitoneal shunting was required 9 days postoperatively because of persistently elevated ICP. The other seven patients with direct surgery had successful cyst excisions without intraoperative or postoperative complications, and remained asymptomatic without recurrent com-

promise of cerebrospinal fluid (CSF) circulation on postoperative evaluation periods with a mean of 32 months.

In one patient a 2-cm cystic lesion migrated from the third to the fourth ventricle during completion of his preoperative evaluation.

Aqueductal Infestation

Four patients who presented with chronic or subacute headache and signs of increased ICP manifested focal aqueductal stenosis. Evidence of cysticercosis infestation was obtained from a combination of immunological, soft-tissue plain film radiographic, and brain CT assessments. These patients were successfully treated with VP shunting procedures (Table 1). Mean follow-up periods have now exceeded 34 months. It is notable that at no time has intraventricular contrast enhancement been observed in this group of patients.

Fourth Ventricle Cysts

Twenty-four cases were treated for isolated cystic involvement of the fourth ventricle (Table 1). Presentation was predominantly that of chronically increased ICP for periods ranging from 6 weeks to 14 months. However, 11 patients had acutely progressive courses which prompted emergency admission and urgent ventriculostomy.

Seven patients who presented with increased ICP and no signs of focal compression within the posterior fossa underwent primary VP shunting. Three of these later required fourth ventricular exploration for cyst expansion and the development of focal compression signs at 8, 11, and 48 months after the initial procedures.

Seventeen patients had primary excision of cystic lesions limited to the fourth ventricle: 35% of these cysts were of the racemose form. Three of the excised lesions had demonstrated pericystic contrast enhancement on CT scanning, and cyst excision was incomplete in each of these cases because of severe ependymitis. This group, as well as three other patients who had no contrast enhancement but evidence of ependymitis at surgery, eventually required secondary VP shunting procedures at 4, 5, 7, 9, 74, and 102 days following the primary procedure. None of the 11 patients without evidence of ependymitis required CSF diversion during follow-up periods with a mean of 38 months.

Illustrative Cases

On review of this series certain problems and considerations germane to patient presentation, lesion peculiarity, and surgical management became evident.

Local Cyst Expansion and Pericystic Contrast Enhancement

Three cystic lesions increased in volume, causing local mass effect which made direct approach for local decompression ultimately necessary in cases where treatment with CSF diversion was the initial management. Local intraventricular and pericystic contrast enhancement on CT scanning has indicated adhesive ependymitis, and has predicted difficulty in realization of total cyst removal as well as the need for CSF shunting.

Case 1. This 40-year-old woman from El Salvador underwent evaluation for chronic headache of 5 months' duration. A CT scan demonstrated generalized ventriculomegaly without contrast enhancement. A 1-cm fourth ventricular cystic lesion was apparent on contrast ventriculography (Fig. 1). A VP shunt was placed with resolution of her headache.

Six months later she presented with headache and increasing obtundation. Shunt revision was required. Two months later she returned with dysconjugate gaze, nystagmus, and right-sided dysmetria, and a CT scan demonstrated enlargement of the fourth ventricular cyst with complete obliteration of the contours of the fourth ventricle and pericystic enhancement (Fig. 2). On posterior fossa craniectomy, adhesive ependymitis was associated with a 3.5-cm fourth ventricular cystic lesion which was adherent over 60% of its surface to the floor and lateral recess. Removal of the cyst wall was incomplete, but decompression was effected. Disappearance of dysmetria and nystagmus was nearly immediate, but complete resolution of dysconjugate gaze was not realized and persists in stable form 6 months following decompression.

Acute Clinical Progression of Third Ventricle Cyst

Acute clinical deterioration has been associated with cystic involvement in all ventricular compartments; in fact, nearly 40% of these patients had acute progression over periods of 2 to 36 hours. Posterior third ventricular cystic lesions with minimal ventriculomegaly may present technical problems in excision. These may be approached through a transcalsalos interforniceal corridor which allows for initiation of alternative drainage access and complete exploration of the third ventricle in cases of minimal ventriculomegaly.

Case 2. This 20-year-old Mexican male gardener presented with the acute onset of frontal headache and progressive lethargy over a 4-hour period. No focal neurological findings or papilledema was apparent on admission. Emergency CT scanning demonstrated moderate hydrocephalus, and metrizamide ventriculography showed a well circumscribed mass measuring 1.5 cm in size at the iter orifice (Fig. 3). Ventricular fluid contained 12 monocytes, a protein level of 42 mg%, and a cysticercosis titer of 1:8. Clinical improvement ensued after ventriculostomy and, 48 hours following admission, the patient underwent transcalsalos interforniceal excision of a posterior 1.5-cm cystic lesion. This was effected with a 2.5-cm incision in the corpus callosum, fenestration of the septum pellucidum, and a 2-cm incision of the fornical raphe midline. Although the foramina of Monro were visualized, the cyst was not appreciated until the diencephalic roof.
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Fig. 1. Case 1. Metrizamide ventriculogram showing a 1-cm filling defect at the outlet of the fourth ventricle consistent with a cysticercosis cyst (arrows).

was opened and the internal cerebral veins retracted. The lesion was then accessible and was removed with minimal dissection of the tela choroidea. Postoperative recovery was uneventful. The patient has returned to his previous occupation and is without complaints after 27 months of evaluation.

Cyst Migration

Considerations for surgical therapy and preoperative management must be appraised bearing in mind the danger of cyst migration and associated clinical phenomena and potential surgical pitfalls. The following case of cyst migration through the aqueduct of Sylvius demonstrates the possibility of exploration of the wrong ventricular component in an effort to provide surgical decompression.

Case 3. This 12-year-old Mexican-born boy was admitted with a 2-week history of headache, nausea, and vomiting. Bilateral papilledema was apparent on physical examination. A CT scan demonstrated lateral and third ventriculomegaly and a normal-sized fourth ventricle (Fig. 4A). Contrast ventriculography via a left frontal ventriculostomy showed a third ventricular filling defect (Fig. 4B). The evening prior to planned transcallosal third ventricular exploration the patient developed a decreased level of consciousness, Cheyne-

Fig. 2. Case 1. Computerized tomography scans obtained 8 months following the initial ventriculogram in Fig. 1. Left: Plain scan showing total obliteration of the fourth ventricle. Right: Following the intravenous infusion of contrast material a large ring-like enhancement is seen in the region of the fourth ventricle.

Fig. 3. Case 2. Left: Metrizamide ventriculogram showing a 1.5-cm cyst in the posterior region of the third ventricle. The massa intermedia lies anterior to the cyst. Right: Computerized tomography scan following the ventriculogram again demonstrates the cyst in the posterior third ventricle. There is an air bubble in the left frontal horn.

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Stokes respiration, and decerebrate posturing; a right frontal ventriculostomy was placed with subsequent resolution of the patient’s signs. A CT scan demonstrated normal third ventricular volume and contour (Fig. 4C); a metrizamide ventriculogram now revealed the cyst in the fourth ventricular cavity (Fig. 4D). A posterior fossa craniectomy was performed and the mobile cyst was removed intact by gentle aspiration. Postoperative recovery has been complete and uneventful at 11 months.

**CT-Guided Stereotaxic Endoscopic Excision of a Mobile Cyst**

Mobility of cystic elements presents a threat related to acute obstruction of ventricular channels. Stereotaxic CT-guided endoscopic excision of such lesions conceptually offers a plausible remedy.

**Case 4.** This 37-year-old Mexican-American man presented with acute headache, onset of coma, bilateral extensor posturing, and a dilated and partially reactive left pupil. Endotracheal intubation, hyperventilation, and osmotic diuretics were instituted, and a CT scan demonstrated bilateral ventricular enlargement, greater on the left than the right (Fig. 5A). Ventriculostomy was followed by contrast ventriculography which showed a 2.5-cm cystic lesion originally located at the foramen of Monro in the left lateral ventricle, but freely mobile within that lateral ventricular space (Fig. 5B and C). With postural manipulation the lesion was localized and maintained in the occipital horn (Fig. 6).

Following the administration of high-potency glucocorticoids, the patient was placed in a Brown-Roberts-Wells stereotaxic system under local anesthesia. The cystic lesion was readily excised with a flexible forceps through a 6.8-mm endoscope instrument. Cyst rupture occurred during extraction. Throughout the course of this excision the ventricular system was flushed with Ringer’s lactate solution through the endoscopic instrument. The patient had a transient low-grade febrile response and slight headache, but no significant adverse response to this mode of therapy. Ventricular size returned to normal and no further therapy has been required during 11 months of evaluation.

**Discussion**

The most common world-wide parasitic infection of the nervous system, cerebral cysticercosis, results when man acts as intermediate host of the pork tapeworm *Taenia solium*. Infestation of tissues with the larval form of the cestode results from ingestion of foods or liquids contaminated with eggs from mature proglottids, or autoinfection in which an individual harboring the adult parasite transfers egg-laden feces from anus to fingers to mouth or from reversed peristalsis that carries mature proglottids from the small bowel to the stomach. Oncospheres penetrate the wall of the stomach and disseminate systemically through vascular and lymphatic channels. Neurocysticercosis represents the chief manifestation of this dissemination, being responsible for approximately 60% of the symptomatic presentations of this infestation. Intraventricular involvement is apparent in 15% to 20% of those cases with neural compartment infestation.42

Grossly, the parasite may be appreciated as a discrete encapsulated cystic form that contains a recognizable portion of the parasite or as a delicate thin-walled vesicular racemose form that commonly contains no recognizable scolex of the cestode. The cysticercosis of *Taenia solium* in biopsied specimens is not difficult to distinguish from the larger cysts of *Echinococcus granulosus*. It contains a single invaginated scolex, which is characterized by four suckers and a double row of hooklets. Cysticerci of *Taenia saginata* are not found in the human brain. The only tapeworm larva that may
cause confusion is that of *Multiceps multiceps* from the dog. This does not produce a cysticercus with a single scolex, but a coenurid larva containing a number of scolices.

It is considered that the oncosphere reaches the ventricular cavity by way of the choroid plexus; gaining entry, cyst formation may impact upon the normal anatomical structures by a number of mechanisms. Cystic lesions harboring viable parasites may at various stages of volume development migrate through the ventricular system, occluding vital communication corridors and initiating acute episodes of ventriculomegaly with sudden death, or causing mass effect with focal compression of neural elements and local neurological findings. Larval death initiates permeability of the cyst wall causing either focal or generalized ependymitis with the potential for occlusion of ventricular outlets.  

As in our observations and the statistical evaluations of others, transventricular migration is implied toward the fourth ventricle where phenomena related to the presence of such cysts is most often evident.\(^{52,53}\)

Diagnosis may be suspected and largely verified by combinations of data and analytical assessment of factors related to 1) individual exposure to an endemic area; 2) CT findings; 3) peripheral blood or CSF eosinophilia; 4) CSF lymphocytosis; 5) immunoassays of blood or CSF; and 6) evidence of other organ involvement.

Histological assessment of cystic components is essential for absolute diagnosis, as serum and CSF diagnostic techniques are neither consistently helpful nor reliable. Eosinophilia is periodic and nonspecific. Serum indirect hemagglutination titers to cysticercal antigenic products may be employed, but do not offer absolute evidence either for or against infestation.\(^{45}\) Indirect immunofluorescent antibody and agar gel precipitin assays are unreliable indices of specific infestation.\(^{48}\) Cerebrospinal fluid assessment may be "suggestive," but lymphocytic pleocytosis and elevated protein content are nonspecific indicators; complement fixation or precipitation assays are supportive, but not absolute tests for this disorder.

The anatomical substrate and its pathological structural alterations are defined by plain and contrast-enhanced CT scans as well as contrast ventriculography. In nearly all cases, positive-contrast ventriculography, with or without CT scanning, is essential for identification of cystic presence and definition of the lesion's extent and contour. Ventricular and periventricular contrast enhancement has indicated ependymitis, with implications of difficulty in accomplishing complete surgical cyst excisions, and suggesting the need for shunting and other means of CSF diversion. It is important to consider that ependymal alterations may be present in the absence of such enhancement and may
reflect a quiescent and later stage of the response. As the major manifestations of intraventricular infestations are cystic development or ependymal responses that may be either focal or generalized, it has been stressed that compartmental ventriculitis may be present and evolve as a "double-compartment" hydrocephalic process.17 This phenomenon has been observed particularly within the fourth ventricle. Although we have observed this process in conjunction with cisternal and basal arachnoiditis, we have not encountered it as an isolated finding.

In selecting the surgical approach, the surgeon should consider the possibility of ependymitis which may be associated with cystic lesions. Recognizing the potential for concurrent or eventual outlet obstruction, transcortical corridors and fenestrations of the septum pellucidum have been established to offer alternative drainage pathways.33,36 Successful primary direct surgical management of solitary cystic lesions was realized in all the 21 patients in this series who demonstrated no ependymitis or evidence of associated focal ventricular obstruction at surgery. Mean time of postoperative assessment in this group exceeds 36 months.

The potential for cyst migration must be considered in preoperative assessment, as postural changes may initiate changes in cyst location within a given ventricular cavity, or frank transit from one ventricular cavity to another, as noted in Case 3. The data accrued in these patients argue for direct surgical excision of cystic lesions as their location or migration may produce not only chronic, but also acute and possibly lethal clinical progression. In addition, local mass effect, as noted in Case 1 and described by others,37 may evolve in cases where indirect therapy is employed.

Although ventriculostomy may be indicated, it is apparent from this series that permanent CSF diversionary procedures39 are not necessary therapeutic adjuncs in management of the entire spectrum of intraventricular cysticercosis. Direct surgical excision of simple cystic lesions,55 in the absence of radiographic or surgical evidence of arachnoiditis or ependymitis, appears to be an adequate primary therapeutic strategy in the majority of cases.

Current surgical techniques allow for safe and accurate access to virtually any intracranial point.3 Computerized tomography-guided stereotaxis combined with appropriate endoscopic instrumentation offers a potential technique for cyst excision in the lateral and third ventricular regions. Case 4 illustrates this concept which merits consideration, certainly for lateral ventricular lesions with no evidence of surrounding contrast enhancement. Although certain authors40 have considered cyst rupture a dangerous event, others34,37 would not share this view. Our experience with Case 4 and with four other open procedures with associated cyst rupture would lend further support to the latter viewpoint. In all cases, high-potency glucocorticoids and vigorous intraventricular lavage with body-temperature Ringer's lactate solutions were employed.

A striking feature of our clinical experience has been the high incidence (38%) of patients presenting with rapid clinical deterioration; in this group, symptomatic predictors of the processes were either minimal or non-existent. The nature of this disorder, combined with consideration for the social character of the immigrant groups affected, calls for an increased awareness of the potential for the presentation of the problem within these populations.

The role of the agent praziquantel,† which is currently being employed by our Mexican colleagues and in early trials in the United States, in relation to the overall management of this disorder will need to be determined over the next decade. It is hoped that the compound will prove to be effective, particularly in parenchymatous involvement, within acceptable limits of toxicity.

Conclusions

Our experience with 45 cases of intraventricular cysticercosis indicates that patient management and operative planning and expectation should be influenced by consideration of: 1) the potential for sudden death; 2) the potential for cyst migration; 3) associated ependymitis, defined by CT or verified surgically; 4) the potential for increases in cyst volume with local mass effect phenomena; 5) selection and institution of surgical approaches that establish alternative routes of CSF flow; and 6) stereotaxic endoscopic cyst excision.

References


† Praziquantel, which is still in the experimental stage, is manufactured by E. M. Industries, 5 Skyline Drive, Hawthorne, New York.
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