Hydrocephalus in cerebral cysticercosis

Pathogenic and therapeutic considerations

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The cases of 11 patients with hydrocephalus secondary to cerebral cysticercosis are analyzed. Most of the patients had suffered from epilepsy before they developed hydrocephalic symptoms, and computerized tomography showed that infestation of the parenchyma coexisted with ventricular or cisternal colonization. In four cases, the parasitic vesicles compromised cerebrospinal fluid (CSF) flow in the ventricular system, resulting in internal hydrocephalus. Communicating hydrocephalus, caused by the presence of Cysticercus larvae in the basal cisterns (Cysticercus racemosus), or by the occurrence of a chronic basal meningitis, or both, developed in seven more patients. Changes in CSF pressure were related to the number and location of the cysts and to the leptomeningeal inflammatory reactions evoked by them. The majority of patients presented with a chronic and relatively normotensive hydrocephalus.

All patients except one had identifiable ventricular or cisternal Cysticercus larvae; these patients were treated with open removal of the cysts, and did well. However, most of them had impairment of CSF flow through the basal cisterns and required permanent CSF shunting. Communicating hydrocephalus due to leptomeningeal scarring was also successfully managed with extracranial shunting. Epilepsy was controlled with anticonvulsant therapy. Although good lasting results may be obtained with aggressive treatment of neurocysticercosis, patients are liable to relapse because surgery is only palliative in most instances.

Key Words: cyst • cysticercosis cerebri • hydrocephalus • cerebrospinal fluid • intracranial pressure • computerized tomography

Cerebral cysticercosis results when man acts as the intermediate host of the pork tapeworm, Taenia solium. After ingestion by the host of contaminated food or water, the ova lose their shell in the stomach, traverse the intestinal wall, and gain access to the bloodstream. Most often they lodge in the nervous system and its coverings, or in the eye, subcutaneous tissues, or skeletal muscles. In a few weeks, the embryo develops a cystic covering. The cyst itself, containing the scolex, is designated "Cysticercus cellulosae." Infestation of the nervous system may cause intracranial hypertension, epilepsy, organic dementia, pseudomeningeal reactions, and symptoms due to compression of the cranial nerves, spinal cord, or cauda equina. Frequently, a combination of these symptoms is observed in a given patient, and the clinical picture depends on the number of parasitic vesicles (from one to more than 200 in a single brain), their size (from pinpoint vesicles found in miliary forms, which appear mainly in small children, to giant lesions measuring more than 6 cm in diameter), the stage of their development (alive or dead), and their anatomic location (parenchymatous, ventricular, or meningeal).

Parenchymatous cysticercosis may be completely asymptomatic, but is usually associated with epilepsy and runs a chronic course. Conversely, colonization of the ventricular system and the basal cisterns of the brain gives rise to quite complex clinical syndromes and carries a poorer prognosis, mainly because the cerebrospinal fluid (CSF) flow is disturbed by different mechanisms. In this paper we analyze a series of 11 cases of hydrocephalus secondary to cerebral cysticercosis.
Clinical Material and Methods

The 11 cases reported here represent nearly half of the total series of patients with neurocysticercosis seen in our hospital in a 7-year period (1974 to 1980). The remainder of the cases had purely parenchymatous infestation and presented with epilepsy but without hydrocephalus. All these patients had lived for a time in certain areas of Spain in which cysticercosis was still endemic.

Apart from the routine laboratory tests, most patients underwent electroencephalography, brain scintiscanning, CSF analysis, and Mantoux tuberculosis and venereal disease tests. All patients, except two who were admitted before computerized tomography (CT) became available, had plain and contrast-enhanced CT scans. Four patients had angiographic and/or ventriculographic studies. After cerebral cysticercosis was found or suspected, x-ray films of the abdomen and extremities were performed to search for systemic infestation.

Case Reports

Internal Obstructive Hydrocephalus

Case 1. This 42-year-old woman had suffered episodes of generalized convulsions for the past 12 years. In 1974, 2 months before hospitalization, she showed subtle behavioral changes and defective short-term memory. Three days before admission, she complained of intermittent headaches and became lethargic. At examination, she was drowsy and showed signs of meningeal irritation. Her CSF pressure was 46 mm Hg, and the analysis was normal. Common carotid angiography disclosed a left temporal avascular mass (Fig. 1). At surgery, the temporal lobe bulged due to dilatation of the temporal ventricular horn, which was isolated from the atrium by a thin-walled translucent cyst, 1 cm in diameter. The cyst, which floated freely in the CSF, was removed without spillage of its contents, and histopathological study showed it to be a cysticercus. At last examination, 6 years after surgery, the patient was in good condition and we found no evidence of recurrence.

Case 2. This 38-year-old man had suffered intermittent headaches, and memory and behavioral disturbances for 5 months before admission in 1980. Admission funduscopy was normal, and there was generalized hypertonus. His memory for recent events and judgment was impaired. His gait was unstable, with a wide base and short steps. A CT scan disclosed asymmetrical dilatation of the lateral ventricles, and normal-sized third and fourth ventricles (Fig. 2). Sev-
eral small, round calcifications were seen scattered through the cerebral hemispheres. Small calcified densities compatible with cysticercosis were also seen in the soft tissue. A diagnosis was made of systemic cysticercosis with bilateral ventricular hydrocephalus, probably due to secondary ependymitis. The patient's family refused metrizamide ventriculography and treatment.

Case 3. This 55-year-old man showed insidious behavioral changes with defects in memory for 18 months before admission in 1980. Six months before admission he developed gait disturbances and complained of episodic headaches. On examination, he appeared disoriented and apathetic, and exhibited disorders of memory and judgment. There was generalized hypertonus, and the gait was apraxic. Bilateral papilledema and left-sided dysmetria were also observed. A CT scan showed supratentorial hydrocephalus, and multiple areas with a density similar to that of CSF were seen in the left cerebellar hemisphere and the suprasellar and frontal interhemispheric regions (Fig. 3). Multiple small areas of calcification were identified in the skeletal muscles. The CSF pressure, which was continuously measured before placement of a ventriculoperitoneal shunt, ranged between 19 and 48 mm Hg. Lumbar CSF analysis revealed 46 cells/ml (30% lymphocytes), 30 mg/100 ml glucose, and 140 mg/100 ml protein.

After the shunt was placed, the patient's papilledema subsided and his mental defects cleared completely. Although his gait was much improved, it was still unstable, and upward conjugate gaze was limited. A posterior fossa craniectomy was performed, and a

Fig. 3. Case 3. Preoperative computerized tomography scans showing hypodense lesions in the left cerebellar hemisphere and in the incisural, suprasellar, and frontal interhemispheric regions. There is increased periventricular lucency. A small area of calcification at the tip of the left frontal lobe can also be seen (arrow).

Fig. 4. Case 4. Left: Pantopaque ventriculogram showing supratentorial hydrocephalus and a dilated aqueduct caused by a mass in the fourth ventricle. Right: Cysticercus cysts removed from the fourth ventricle.
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cluster of thin-walled cysts occupying the superomedial part of the left cerebellar hemisphere was removed. Because of their parenchymatous location, none of the cysts could be removed without spillage of their contents. After this operation, signs of brainstem compression disappeared. The cysts were diagnosed as cysticercus.

Case 4. Nine months before admission in 1974, this 54-year-old woman suffered episodes of violent headaches associated with movements of the head and accompanied by dizziness and vomiting. Seven months later, she noticed a decrease in visual acuity, and developed truncal ataxia. At examination, bilateral papilledema and optic atrophy were noted. Her gait was ataxic. X-ray films of the chest and extremities showed rod-shaped calcifications in the skeletal muscles. Skull films indicated chronically elevated intracranial pressure, and carotid angiography revealed hydrocephalus. Pantopaque ventriculography disclosed a dilated aqueduct and a mass occupying the fourth ventricle (Fig. 4 left). Mean ventricular fluid pressure was 50 mm Hg, and CSF analysis yielded 20 cells/ml (90% lymphocytes) and 40 mg/100 ml protein.

A posterior fossa craniectomy was performed, and eight cysts were removed from the fourth ventricle. Three cysts, which were joined by a common pedicle, were translucent; they measured 0.5 to 1 cm in diameter. The other five were opaque, containing a jelly-like material (Fig. 4 right). After removal of the cysts, the CSF flowed freely through the aqueduct. Histopathological study of the cysts showed them to be Cysticercus larvae, some of which had degenerated and contained calcium deposits. The postoperative course was without complications; however, the patient remained bedridden with mental disorders and urinary incontinence. The ventricular enlargement did not clear, although postoperative CSF pressure was within normal limits. A ventriculostomy shunt was then placed, and the patient's mental and gait disturbances improved until she reached an almost normal state. In the last 5 years she has had two shunt revisions. A control CT scan showed three small calcifications in the brain.

Communicating Hydrocephalus and Cysticercus Racemosus of the Basal Cisterns

Case 5. This 56-year-old woman had had infestation with tapeworm in childhood. At the age of 24 years, she began to have focal and generalized ictal attacks. One year before admission in 1975, she was operated on for cauda equina cysticercosis, and 6 months later she started to complain of headaches and disturbances of short-term memory. Funduscopic examination disclosed ventricular enlargement and basal hypodense lesions. Funduscopic examination disclosed ventricular enlargement and basal hypodense lesions compatible with cysticercus (Fig. 5 lower). Mean CSF pressure was 7.4 mm Hg, but ranged between 1 and 18 mm Hg. Analysis of the CSF yielded 34 cells/ml (60% lymphocytes) with 34 mg/100 ml glucose and 110 mg/100 ml protein. In a single surgical session, the basal cysticercus cysts were removed and a ventriculoperitoneal shunt was placed. After this second operation, the patient was much improved. A control CT scan showed the disappearance of hydrocephalus and the absence of cysts.

Case 7. This 53-year-old man had suffered episodes of generalized convulsions from the age of 25 years. For more than 5 months before admission in 1979, he complained of headaches; he had lost the control of sphincter muscles and exhibited mental changes with
Fro. 5. Case 6. Upper: Initial computerized tomography (CT) scans show multiple oval and round hypodense areas located in the basal cisterns, with associated hydrocephalus and a great hypodense mass in the left ventricular atrium. Lower: Metrizamide-enhanced CT scan obtained 9 months after the first operation showing the return of hydrocephalus and the presence of basal cysts, which appear as unenhanced masses.

Fro. 6. Case 7. Left Pair: Preoperative computerized tomography (CT) scans showing ventricular enlargement and increased periventricular lucency. A small, rounded calcification is seen at the tip of the left occipital lobe (arrow). Hypodense areas in the Sylvian fissures simulate brain atrophy. Right Pair: After shunting, hydrocephalus disappeared, but the Sylvian lesions appear to be increased in size and medially displaced. The intravenous injection of contrast material did not change the appearance of the lesions.

defects in memory, and progressive gait disturbance. This clinical syndrome fluctuated episodically, on several occasions reaching almost complete regression. At the referring hospital, he was suspected of having tuberculous meningitis because a CSF analysis yielded 106 cells/ml (95% lymphocytes), 30 mg/100 ml glucose, and 80 mg/100 ml protein. At admission, the patient appeared akinetic and was mute. Generalized hypertonus and extensor plantar responses were noted. Funduscopy was normal. Multiple calcifications resembling grains of rice were seen in the skeletal muscles of the legs. A brain CT scan disclosed generalized ventricular enlargement, and irregular areas with a density similar to that of CSF were seen in the Sylvian fissures and the basal cisterns (Fig. 6 left pair). Small calcifications, which were not visualized in the plain films of the skull, were also seen in the cerebral hemispheres. The CSF pressure was measured before placement of a ventriculoperitoneal shunt; it ranged between 10 and 30 mm Hg, with a mean of 14.7 mm Hg. Analysis of CSF was normal at this time. Immediately after the operation, the patient regained the ability to speak and stand. Control CT showed a decrease in the ventricular size, but the hypodense lesions in the Sylvian fissures appeared to be larger and more medially located than before shunting. Unexpectedly, the patient's neurological status deteriorated until he became comatose. A repeat CT scan showed complete regression of hydrocephalus (Fig. 6 right pair), but the patient did not improve and died before the basal lesions could be dealt with.

Chronic Communicating Hydrocephalus Secondary to Basal Arachnoiditis

Because of their similar clinicopathological presentation, Cases 8 to 11 have been summarized in
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Table 1. The radiological features in these cases are shown in Fig. 7.

Discussion

Ventricular cysticercosis may cause internal obstructive hydrocephalus by different mechanisms. After the oncosphere reaches the ventricular lumen by way of the choroid plexus, it forms a cyst which may run different courses. While the encysted larva is alive, it grows steadily without producing a reaction from the host, but can mechanically interfere with CSF flow. Cysts can migrate along the ventricular cavities, and the sudden impact of a vesicle in the ventricular foramina or the aqueduct may cause acute hydrocephalus and the death of the patient. When the larva dies, the cyst wall becomes permeable, and its toxic contents provoke an inflammatory reaction liable to cause granular ependymitis, which may also occlude the ventricular or the aqueductal lumen.

Our Case 1 demonstrates the production of unilateral ventricular hydrocephalus by a single cysticercus which caused the dilatation of the temporal horn. Obstruction was due merely to the presence of the cyst which floated in the ventricular fluid. Intraventricular cysticerci are solitary in 7% of cases, and the most frequent location is in the fourth ventricle. In Case 2, obstruction of the foramina of Monro, which was probably due to secondary ependymitis, resulted in bilateral ventricular hydrocephalus. A similar case, in which extensive adhesions in the anterior third ventricle were observed at surgery, has been reported recently. In Case 3, supratentorial hydrocephalus resulted from the extrinsic compression of the aqueduct by a cluster of cysts located in the cerebellum. Finally, in Case 4, supratentorial hydrocephalus and dilatation of the aqueduct were due to fourth ventricle cysticercosis. Removal of the cysts alleviated intracranial hypertension in this patient. However, ventricular enlargement persisted for several weeks until a CSF shunt was placed. Communicating hydrocephalus was probably caused by a concomitant basal arachnoiditis in this case.

When cysticerci are small enough to traverse the foramina of the fourth ventricle, they reach the basal cisterns and proliferate, giving rise to the racemose forms that usually result in generalized ventricular enlargement. Dying cysts, by evoking inflammatory reactions, may cause chronic basal meningitis which also results in communicating hydrocephalus. Al-

| TABLE 1 |
|-----------------|-----------------|-----------------|-----------------|-----------------|
| Major findings in four patients with basal arachnoiditis and communicating hydrocephalus* |
| Factor | Case 8 | Case 9 | Case 10 | Case 11 |
| age (yrs) | 52 | 51 | 49 | 57 |
| sex | F | F | M | F |
| history | tapeworm infestation; gen seizures & headaches, 5 yrs | gen seizures, 26 yrs | gen seizures, 30 yrs; recent headaches & gait instability, 5 yrs | gen seizures, 25 yrs |
| recent symptoms | dementia, depression & gait apraxia, 9 mos; incontinence, 3 mos | headaches, dementia & gait apraxia, 3 mos; incontinence, 2 mos | gait apraxia & memory defects, 5 mos | dementia & gait unsteadiness, 9 mos; headaches, 3 days; coma, 1 day |
| CSF pressure range (mm Hg) | 6-30 | 12-27 | 7-32 | 5-35 |
| mean (mm Hg) | 10 | 14.4 | 8.6 | 10.5 |
| CSF analysis | cells/ml | 80 | 100 | 68 | 19 |
| % lymphocytes | 100 | 20 | 90 | 8 |
| protein (mg%) | 100 | 100 | 49 | 100 |
| glucose (mg%) | 68 | 80 | 22 | 51 |
| x-ray study skull calcifications | normal | normal | normal | normal |
| soft tissue calcifications | not done | calcifications | ruled out ventricular cysts | not done |
| PEG findings gen hydrocephalus; small calcific densities | no change | gen hydrocephalus; small calcific densities | no change | gen hydrocephalus; small calcific densities |
| CT scanning plain enhanced treatment result | VA shunt, good: 2 episodes of meningeval reactions, 2 shunt revisions | VA shunt, excellent: 2 episodes of meningeval reactions, 3 shunt revisions | VP shunt, excellent | VP shunt, excellent: 2 episodes of meningeval reactions, 1 shunt revision |
| follow-up period (yrs) | 6 | 3 | 1 | 2.5 |

* Abbreviations: CSF = cerebrospinal fluid; PEG = pneumoencephalogram; CT = computerized tomography; VA = ventriculoatrial; VP = ventriculoperitoneal; gen = generalized.
FIG. 7. Upper: Case 10. Enhanced preoperative computerized tomography (CT) scan showing generalized hydrocephalus as well as living and dead larvae in the cerebral hemispheres. A ventriculographic study ruled out the presence of Cysticercus cysts in the ventricles. Lower: Cases 8 (D), 9 (E), and 11 (F). Preoperative CT scans showing ventricular dilatation with increased periventricular lucency and dead cysticerci in the parenchyma. There were no changes in these cases following contrast enhancement.

Although these two pathological processes may occur simultaneously or follow each other in a given case, we have separated our seven patients with communicating hydrocephalus into two groups on the basis of the dominant underlying pathology. In Cases 5, 6, and 7, generalized hydrocephalus was associated with basal cistern colonization by Cysticercus racemosus. In Case 5, we directly attacked the basal cysts and, despite the previous occurrence of meningeal reactions, the patient did not develop further mental or gait disturbances and hydrocephalus was arrested. Such an outcome is unusual, and the possibility of a relapse cannot be dismissed in this case. In Case 6, the only patient with mixed intraventricular and cisternal cysticercosis, all the cysts were removed at a single operation. The initial postoperative course was fairly good, but 9 months later the patient developed mental and gait disturbances that resolved with permanent CSF shunting. This case clearly shows that relapse may occur a few months after an apparent complete removal of a basal Cysticercus racemosus. Because of the impossibility of removing all the basal cysts at a single operation, we initially placed an external shunt in Case 7. Immediately after shunting, the patient was much improved, but unexpectedly became comatose and died before we could deal with the basal cysts. This case illustrates the fact that CSF shunting alone is worthless in the patient with massive basal cysticercosis. Cases 8 to 11 had generalized hydrocephalus, apparently in the absence of healthy cysticerci in the basal cisterns. Thus, most likely they suffered from chronic basal meningitis. In fact, these patients’ CSF analyses showed changes indicative of meningeal irritation, and they had suffered previous recurrent episodes of meningitis, lasting for several days, then subsiding for a while and reappearing after a variable period. Finally, a chronic low-pressure hydrocephalus resulted. Although these four patients were much improved by CSF shunting, and control CT scans showed the disappearance of hydrocephalus, three of them have had recurrent pseudo-meningeal reactions similar to those they displayed before shunting, indicating that the disease is still active.
Before the advent of the CT scanner, the diagnosis of ventriculocisternal cysticercosis was difficult because the angiographic and ventriculographic findings were nonspecific. With CT scanning, both the living encysted and the dead calcified larvae may be clearly visualized wherever their location, and the degree and type of hydrocephalus is also easily ascertained. Although in our experience the intravenous contrast material did not seem to change the attenuation values of the cysts and the surrounding brain, both a pericystic and basal cisternal enhancement have recently been reported by other authors. Since the living larvae have absorption values similar to CSF, positive contrast studies or a metrizamide-enhanced CT scan are necessary for an accurate visualization of the cysts masked within the enlarged CSF spaces.

Excellent results or even a cure may ensue after open surgery to remove intraventricular Cysticercus cysts when they have not yet evoked ependymal or leptomeningeal reactions. Although the prognosis in the basal racemose forms has been reported as very poor, in the past, surgery was unrealistic and incomplete in many cases because the neuroradiological studies failed to show the precise location and number of the cysts. With the information provided by the CT scan, the approach to a basal racemose form should be planned to attack mainly those cysts that obstruct CSF flow, as there are many cysts in other parts of the brain that are completely asymptomatic. If possible, both intraventricular and cisternal cysts should be removed without spillage of their contents. However, the risks from cyst rupture have been minimized by Loyo, et al., and the absence of postoperative complications in our Cases 3 to 6 supports these authors’ statement. Extracranial CSF shunting is the only efficient treatment for cases of communicating hydrocephalus secondary to chronic basal meningitis. This form of therapy, rather than ventriculocisternostomy, is also advised in patients with internal obstructive hydrocephalus due to granular ependymitis, because some degree of basal arachnoiditis is frequently associated in these cases.

There is no effective specific medical treatment for cerebral cysticercosis. High-dose corticotherapy may be of great help in the management of the pseudomeningeal or pseudotumoral reactions caused by basal and parenchymatous cysts, respectively. Seizures are easily controlled with anticonvulsant therapy in most cases.

References
20. Trelles JO, Lazarte J: Cisticercosis cerebral; estudio clinico, histopatologico y parasitológico. Rev Neuropsiquiatr 3:393–511, 1940