Contemporary Neurosurgical Approaches to Neurocysticercosis

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Abstract. Neurocysticercosis (NCC) includes a wide spectrum of illnesses. Newer neurosurgical approaches are being applied to NCC. Although clinical trials are leading to a consensus on medical management, fewer data address neurosurgical approaches. We reviewed all neurocysticercosis patients evaluated by neurosurgery at Ben Taub General Hospital, Houston, Texas, between August 1997 and December 2005. From 31 patients that had a neurosurgical evaluation, 16 patients were treated with shunts (5 had shunt failure requiring revision), 13 by surgical resection of cysts (9 underwent craniotomy and 4 endoscopy), and 2 by medical therapy. A fifth endoscopy was performed in one patient with shunt failure. None of the endoscopic patients needed another intervention. Despite the availability of anti-parasitic and anti-inflammatory therapies, neurosurgery continues to play an important role in the management of selected cases of NCC. In contrast to the high rate of shunt failure, neuroendoscopy seems to be associated with higher success rate than any other neurosurgical approach.

INTRODUCTION

Neurocysticercosis (NCC) is caused by infection of the human central nervous system (CNS) with the larval stage of the parasite *Taenia solium*. NCC is the most frequent cause of seizures and hydrocephalus in adults from regions where the disease is endemic, including Central and South America, Asia, Africa, and Eastern Europe.1–4 NCC has recently become more prevalent in the United States because of the immigration or travel from endemic regions.

NCC produces a wide spectrum of clinical forms depending on the number of parasites, their location, and the degree of host inflammation. Experts agreed that different therapeutic approaches should be undertaken depending on the specific disease presentation. Most patients present with cysticerci in the brain parenchyma and present with seizures. In contrast, extraparenchymal cases are more likely to present with hydrocephalus and are at increased risk of fatal complications. Regardless of the clinical form being treated, symptomatic therapy (i.e., anti-epileptic medications, surgical cerebrospinal fluid [CSF] diversion procedures, and/or corticosteroids) must be the initial goal of management in NCC.5,6

Multiple observational and some randomized studies are defining the optimal medical management of parenchymal NCC. Treatment with cysticidal agents, including albendazole and praziquantel, has been the focus of the majority of these reports.7–10 In contrast, very few data are available regarding surgical options for treatment of NCC, particularly of extraparenchymal cases. No clear consensus can be inferred from the literature, and the optimal management of intraventricular and subarachnoid NCC remains to be elucidated.5

The objective of this paper is to describe current neurosurgical approaches for the treatment of NCC and determine their different outcomes. To accomplish our objectives, we performed a retrospective study of all patients with NCC who were surgically treated at the Department of Neurosurgery, Ben Taub General Hospital (Houston, TX).

MATERIALS AND METHODS

We retrospectively reviewed all inpatient and outpatient charts of patients with NCC who had been surgically treated in the Department of Neurosurgery at Ben Taub General Hospital (Harris County Hospital District), Baylor College of Medicine, from August 1997 through December 2005. Only patients with a definitive or probable diagnosis of NCC by current diagnostic criteria were included.11 Those cases with incomplete information or unavailable charts were excluded. The protocol was approved by the Institutional Review Board or Baylor College of Medicine, Houston, Texas. Classification was done according to the location of the cyst or cysts based on computed tomography (CT) and/or magnetic resonance imaging (MRI) as follows: parenchymal and extraparenchymal, the latter was subdivided in ventricular and subarachnoid forms. Surgical procedures included craniotomy and removal of cysts, ventriculo-peritoneal (VP) shunt and endoscopy (including third ventriculostomy and endoscopic cyst extraction). Outcome was evaluated with clinical and radiographic follow-up (Table 1).

RESULTS

Demographics. A total of 31 subjects were eligible, and all of them were finally included into the study. Sixteen cases (52%) were male. The median age was 37.1 years (range: 22–69 years). All subjects were self-identified as Hispanics, and 14 had been born in Mexico (birthplace data available for 17 cases). The median time elapsed from immigration to the United States through hospital admission was 9 years (range: 0–39 years, data available for 15 cases).

Clinical presentation. Headache was present in 29 (94%) patients, seizures in 5 (16%), nausea and vomiting in 14 (45%), altered mental status in 12 (9%), visual symptoms in 10 (32%), paresis in 5 (16%), sensory deficits in 4 (13%), ataxia in 4 (13%), dizziness in 3 (10%), psychiatric symptoms in 2 (6%), and aphasia in 1 (3%). A history of previous CNS disorders, including seizures, viral meningitis, pseudotumor cerebri, and NCC, was recorded in nine (29%) subjects. One patient was co-infected with HIV. This case has been previously reported.12

Location of the disease based on imaging. CT of the brain was done in all patients at the time of admission. MRI was obtained in 27 (87%) patients on admission or shortly after.
Hydrocephalus and cerebral edema were observed in 27 (87%) and 8 (26%) patients, respectively. According to the location of disease, 4 (13%) patients had parenchymal cysts, 19 (61%) had intraventricular cysts (Figure 1), 6 (19%) had subarachnoid basal cisternal cysts (Figure 2), and 2 (6%) had hydrocephalus without cysts identified (Table 1).

Within the group with intraventricular cysts, six patients also had intraparenchymal lesions visualized in imaging studies. Similarly, within the group with subarachnoid cysts, two patients also had intraventricular cysts, and one also had intraparenchymal cysts (Figure 2). Calcified lesions were seen in eight (26%) patients and arachnoiditis was observed in one patient.

**Medical treatment.** Steroids (dexamethasone or prednisone) were required in 19 (61%) patients because of ventriculitis, perilesional edema, or arachnoiditis. Anti-epileptic drugs (phenytoin or levetiracetam) were used in 15 (48%) patients;
and anti-parasitic drugs (albendazole or praziquantel) were administered in 12 (39%) patients. In most cases, anti-epileptic drugs were administered prophylactically.

**Surgical approaches.** Among all enrolled cases, 16 patients were primarily treated by placement of VP shunts, 13 by surgical resection (9 underwent craniotomy and 4 underwent endoscopic removal of cysts), and 2 by medical therapy (anti-parasitic and anti-inflammatory drugs).

**Parenchymal NCC.** Four patients with parenchymal NCC underwent primary craniotomy and excision of the cysts. The indication for surgery was for the diagnosis of a presumed tumor or abscess.

**Subarachnoid NCC.** Five of six patients underwent primary VP shunt placement and, among those, two of them developed shunt malfunction requiring revision. The remaining patient underwent VP shunt placement during a second hospitalization after failed medical therapy.

**Intraventricular NCC.** Of the 19 patients with intraventricular NCC, 9 were treated primarily by VP shunt placement, 5 by open craniotomy, 4 by endoscopic resection, and 1 by medical therapy. Among the nine treated primarily by shunt placement, there were three documented shunt failures; two of them required shunt revision and the other one needed a craniotomy. Two of the five patients treated by craniotomy developed relapsed hydrocephalus and eventually required placement of VP shunt. The one patient treated medically had a rocky clinical course characterized by seizures and headaches requiring placement of VP shunt and ultimately endoscopic resection of cysticerci. As mentioned above, four patients were primarily treated by endoscopic removal of cysts, and none of them required any additional therapy. An illustrative intraoperative endoscopic image showing removal of a third ventricular cysticercus is shown in Figure 3.

**Hydrocephalus without identifiable lesions.** These two cases were confirmed by a combination of serologic and epidemiologic criteria. Both cases were treated by VP shunt placement; one required a shunt revision.

**DISCUSSION**

Extraparenchymal cysticercosis refers primarily to infection of the subarachnoid space and ventricles. The clinical presentation and treatment differ from parenchymal infection, and generally, the prognosis of these patients is worse. Previous studies reported resolution of large subarachnoid and intraventricular cysts after administration of albendazole, suggesting that anti-helmintic therapy may have some benefit in the treatment of these conditions. Primary medical treatment of extraparenchymal NCC has not gained universal acceptance however. The most persuasive argument against treatment with anti-helmintic therapy alone is that, until the cyst disappears, the patient is still at risk of life-threatening complications. Another consideration is that anti-helmintic treatment accelerates the inflammation process associated with cyst degeneration and can thereby lead to complications such as ependymitis and arachnoiditis, despite the prophylactic use of corticosteroids.

**Parenchymal NCC.** Seizures, mass effect, and focal neurologic signs may be caused by viable or degenerating parenchymal cysts. Medical therapy consisting of anti-convulsants, corticosteroids, and anti-parasitics could potentially accelerate resolution of these lesions and shorten the duration of the symptomatic stage of NCC. In our series, the indication for surgery in these cases was for a suspected tumor or other infection. None were for mass effect. Thus, these procedures may have been avoidable with a higher index of suspicion.

**Subarachnoid NCC.** Subarachnoid NCC is a severe form of NCC associated with a high fatality rate. It may present as large mass-occupying lesions (sometimes termed racemose cysts) or inflammatory responses to subarachnoid cysts (with chronic meningitis, vasculitis, and communicating hydrocephalus). Although medical therapy with anti-parasitic and anti-inflammatory drugs is important, patients still often require surgical approaches to hydrocephalus, as shown by our patient treated medically. Consequently, surgery in subarachnoid NCC is mainly for decompression or VP shunt placement. In our series, the rate of VP shunt failure (two of five patients) remains high. Some have proposed therapy with anti-inflammatory drugs, including methotrexate. Others have argued for prolonged or high-dose anti-parasitic drugs. Whether this can be reduced with aggressive medical therapy with anti-parasitic and anti-inflammatory drugs is unclear. However, even with current management, we noted no fatalities with attention to management of hydrocephalus. Thus, deaths from neurocysticercosis should be avoidable.

**Intraventricular NCC.** Cysts that lodge in the ventricles may result in hydrocephalus by blocking CSF flow mechanically or as a result of inflammation and fibrosis. Blockage of the fourth ventricle and to a lesser extent the third ventricle is relatively frequent and gives rises to the most serious complications.
Although some have advocated a medical approach in these patients, there remains a significant risk for poor outcomes, as shown by our patient initially treated medically. Cyst located in the lateral ventricle can cause unilateral hydrocephalus but may also resolve without sequelae. Hydrocephalus requiring VP shunt is the most common complication of intraventricular NCC requiring treatment. Similar to subarachnoid cases, we observed a high rate (three of eight cases) of VP shunt failure among ventricular cases, which agrees with previous reports. This is thought to be the result of cysts themselves getting sucked into the ventricular catheter or the VP shunt drawing cysts from the posterior fossa into the supratentorial ventricular system. Our endoscopic exploration of ventricles showed the existence, in many cases, of a lacy, mucoid
material surrounding degenerating cysts (Figure 3). We suspect that this material, as well as other inflammation-related debris, could account for some cases of VP shunt obstruction in patients undergoing VP shunt placement without cyst removal. Another potential complication of VP shunt placement without removing a cyst is that deterioration of the cyst can provoke ependymitis and arachnoiditis and thus worsen the prognosis significantly.

Open craniotomy for intraventricular NCC was often complicated by the need for subsequent VP shunt placement in three of seven cases, which makes it a less appealing option. In contrast, neuroendoscopic procedures were performed in five patients, all of whom recovered uneventfully (Figures 1 and 3). Thus, our data suggest significant advantages for endoscopic removal of cysts (intraventricular and perhaps subarachnoid) over the traditional VP shunt placement or open craniotomy.

The transcortical and transcortical-transventricular approaches to the lateral and third ventricle through open craniotomy have been the standard procedures for the removal of intraventricular cysts. Similarly, subfrontal, subtemporal, and pterional approaches are used for removal of cysticercal cysts located in the basal cisterns. These procedures can be technically demanding and are not without the risk of major complications. In addition, transection of the corpus callosum, which is performed during open removal of cysts, can be associated with significant transient memory loss and occasional serious complications including hemiparesis, mutism, and aphasia. In our study, all patients that underwent open craniotomy for removal of cysts had a septum pellucidotomy performed to establish communication between the lateral ventricles to prevent occlusion of the foramen of Monro caused by scarring.

In this report, a rigid endoscope was used to approach the lateral and the third ventricle; however, assessment of the occipital horn and the posterior portion of the third ventricle including the entrance of the cerebral aqueduct were challenging. A flexible endoscope would be more appropriate to explore the entire ventricular system as has been previously reported.

An open microsurgery approach to the posterior portion of the third ventricle has been described, but it is technically demanding, requires a lengthy operative time, and entails significant risk. In contrast, endoscopic navigation through the foramen of Monro arrives at the same location in a matter of minutes, and manipulation of the fornix is not necessary. In addition, it can be possible during the same surgical time to extract cyst from that area.

Endoscopic procedures can also provide the ability to effectively alleviate cases of hydrocephalus. In all our endemic cases, we were able to perform a third ventriculostomy, which resembles the success obtained in adult patients with acquired aqueductal stenosis. Our experience also confirms the observation that the rupture of cysts during endoscopic procedures causes no sequelae, spreading of cysts, or postoperative ventriculitis or arachnoiditis.

In summary, different surgical procedures are still performed in the management of selected cases of NCC and, although endoscopic procedures seem to account for the best outcomes, further studies are still needed to confirm these preliminary observations.