Case Report

Treatment of racemose neurocysticercosis

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Abstract

**Background:** Neurocysticercosis (NCC) is a common parasitic infection of the central nervous system, caused by the tapeworm *Taenia solium*. It is endemic to certain parts of the world, including Central America, South America, Asia, and Africa. The racemose form, characterized by extraparenchymal location, increased morbidity and mortality, and large loculated cystic lesions, is rarely seen in industrialized countries, such as the United States. The management of racemose neurocysticercosis (RNCC) differs from that of the typical parenchymal variant. The ideal course of treatment is debated by experts, but typically includes either surgical intervention with subsequent medical therapy or medical therapy alone.

**Case Description:** We present the case of a 34-year-old male diagnosed with RNCC and treated successfully with surgical cyst drainage, resection, and subsequent medical therapy.

**Conclusion:** Currently, no standardized evidence-based protocol exists that dictate appropriate treatment for extraparenchymal or racemose NCC. We present a case of RNCC treated successfully with surgical and medical intervention. Further research encompassing well-designed clinical trials is necessary to delineate appropriate and standardized protocols for treatment of this disease.

**Key Words:** Cyst rupture, medication, neurocysticercosis, racemose, surgery, *Taenia solium*, treatment

INTRODUCTION

The pork tapeworm *Taenia solium* can cause two different infections in humans: intestinal taeniasis and cysticercosis. The usual life cycle of *T. solium* involves an intermediate host in pigs and a definitive host in humans. The adult stage of *T. solium* infection occurs in humans, resulting in an intestinal tapeworm (intestinal taeniasis) that produces eggs, which once ingested by the intermediate host, are responsible for the larval stage (cysticercosis). The adult tapeworms in humans excrete eggs into the host’s feces, which are subsequently ingested by pigs, developing into cystic forms within the pig’s muscles (cysticerci). Humans ingest the infected

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undercooked pork and the cysticerci develop into adult tapeworms within the human host. In cysticercosis, humans become the accidental intermediate host after ingesting eggs excreted by other humans via a fecal-oral route. After ingestion of the eggs, the oncospheres hatch, penetrate the intestinal wall, and disseminate hematogenously to the muscles, central nervous system, eyes, and other locations.

Central nervous system involvement, known as neurocysticercosis (NCC), occurs frequently in patients with cysticercosis, and is a common cause of new-onset seizures in endemic regions. The tapeworm is endemic in many developing countries, and while not endemic in the United States, it contributes to the burden of disease in patients who present with seizures or intracranial hypertension. Recurrent seizures are the most common presentation, with 30–40% of patients seeking care for epilepsy in endemic countries having evidence of NCC.

Cysts can survive for many years without causing symptoms, but they can degenerate later in life, eliciting a host immune system response and a strong inflammatory reaction. Edema around a degenerating cyst can be interpreted as the inflammatory response of the host towards the parasite. Disease severity and clinical symptoms are dependent on several characteristics of the cyst, including size, number, and location of cysts, as well as host immune response.

Intraparenchymal NCC is the most common form and is most frequently associated with epilepsy. In extraparenchymal NCC, parasitic cysts survive for a variable period of time in the ventricles or subarachnoid space, eventually undergoing a process of involution. This process, however, often results in local inflammation and mass effect. Infrequently, cysts in the subarachnoid space appear as a large cluster of grapes, hence the term “racemose” NCC. Racemose NCC (RNCC) is a less common form of NCC, defined by aberrant proliferation of cestode membrane and arising from segmentation of cysticercosis cellulosae, with the development of new cysts following degeneration of the scolex. They are thus abnormal growths of cystic membranes without a scolex that occur in and around the brain. The subarachnoid (cisternal) form, which includes the racemose variant, is exceedingly rare and even less frequently encountered outside of endemic regions. RNCC is clinically more aggressive than the parenchymal variety and is found primarily in subarachnoid locations.

Clinical manifestations include hydrocephalus, mass effect, chronic meningitis, seizures, cerebral vascular accident, reversible dementia, and cranial neuropathy. The cisternal and ventricular forms of NCC may be missed by CT scans, but can be visualized on MRI with T2-weighted or FLAIR sequences. The 3D MRI sequences, such as the Fast Imaging Employing Steady-state Acquisition (FIESTA) sequence, seem to have the highest sensitivity for identifying the cyst membrane within CSF spaces. Microscopic evaluation of RNCC reveals cysticercal decay with cyst enlargement and hydropic changes without scolecites. The vesicle wall is convoluted with external bulbous projections.

Treatment for parenchymal NCC typically consists of anti-parasitic drugs for viable brain cysts; however, appropriate therapy for RNCC is debated. Some experts have suggested open or endoscopic surgery when possible, with the goal of cyst removal, combined with oral anti-parasitic treatment. In this case, after consultation with two international experts, we elected to offer surgical excision based on the size and the location of the lesion. We believed that surgical excision would provide the best chance for achieving a cure.

CASE REPORT

History/Pertinent findings

A 34-year-old right-handed Hispanic male presented initially to another hospital after a new-onset generalized seizure while driving. He emigrated from Mexico 15 years prior to presentation and has not returned for 6 years. He traveled to many states around the US for work, including Texas, Louisiana, Virginia, Florida, and Nebraska, but has lived in Alabama for the last 11 years. On admission to an outside hospital, he underwent CT head and subsequent MRI brain, which revealed a multi-lobulated cystic lesion of the left fronto-temporal region. A lumbar puncture was performed with cerebral spinal fluid (CSF) labs, culture, cytology, and flow cytometry; all of which were unremarkable. He was evaluated by an outside neurologist and started on oral phenytoin for seizure control. He was subsequently discharged with follow-up at our facility for further management.

Upon evaluation in clinic, he had no focal deficits on neurological examination. Imaging from the another hospital was reviewed. The CT head non-contrast showed a large left fronto-temporal multi-lobulated cystic lesion ~6.5 cm × 4.5 cm × 4 cm in size and primarily centered around the sylvian fissure, causing significant mass effect and 0.5 cm midline shift. Calcifications were present in the central confluence of the cysts. The largest cyst cavity was in the left temporal lobe and did not appear to communicate with the temporal horn. The MRI brain, with and without contrast with T2-weighted sequences, revealed high intensity within the cyst, similar to CSF. FLAIR sequences showed minimal increased signal in the cyst walls. Diffusion-weighted imaging (DWI) sequences showed no increased diffusion restriction. Contrast-enhanced imaging showed no peripheral enhancement of the lesion. Serology for serum cysticercosis IgG antibodies was positive.
Surgical treatment
The patient was treated with dexamethasone 8 mg intravenously every 8 hours on the day prior to surgery and his home phenytoin dose was continued. The CT of his chest, abdomen, and pelvis with contrast were obtained to rule out other possible diagnoses on the differential, including metastatic disease and hydatid cysts; this was unremarkable. The patient was taken to the operating room the following day for surgery and pre-treated with 10 mg of dexamethasone intravenously. A large left pterional craniotomy was performed to sufficiently expose portions of the left frontal and temporal lobes, as well as the sylvian fissure (see supplemental video). Upon opening the dura, discoloration was noted in the superficial pia and arachnoid layers, consistent with chronic inflammation. Stealth navigation was used to plan a trajectory into the anterior and inferior portion of the large temporal cyst and a Touhey needle was inserted through the middle temporal gyrus to aspirate cyst contents, of which 6 mL of clear fluid was obtained. This was performed prior to further dissection to allow for relaxation of the brain. A sylvian dissection was performed, during which the cyst began to evacuate on its own [Figure 2a and b]. Gentle traction was applied to the cyst, which resulted in intra-operative rupture, spilling the cyst contents within the subarachnoid space [Figure 2c]. Copious irrigation was used to clear the cyst fluid and the patient was administered a second dose of 6 mg dexamethasone intravenously. With completion of the sylvian dissection, the cyst complex was removed en bloc with no intra-operative evidence of residual lesion [Figure 2d]. A post-operative neurological exam revealed no neurologic or speech deficits. Non-contrast CT head and MRI brain with and without contrast were obtained on post-operative day 1, which showed expected encephalomalacia surrounding the resection cavity, with no evidence of residual lesion [Figure 3].

Postoperative course
Post-operatively, the patient was started on a 14-day course of oral albendazole 400 mg twice daily. He was discharged from the hospital on post-operative day 5 without complications. He was treated with dexamethasone while undergoing oral anti-parasitic therapy and discharged on a slow steroid taper. He remained on his admission dose of phenytoin at discharge. On scheduled outpatient follow-up, he felt well. Surveillance CT head revealed stable left fronto-temporal encephalomalacia and no evidence of recurrence [Figure 4].

Pathological findings
Neuropathology was consistent with neurocysticercosis in racemose cyst formation [Figure 5]. Grossly, the specimen consisted of tan-white membranous tissue measuring 3.7 cm × 2.5 cm × 0.6 cm. Microscopic examination demonstrated a cyst structure larger than a classic cysticercus cyst, exhibiting multiple convolutions and foldings with rounded protusions at the surface of the tegument. The surface lining exhibited relatively well-preserved microtriches resembling microvilli. Within the inner reticular layer, several small oval or circular calcified bodies (calcareous corpuscles) were present. Scoleces and rostellum (hooklets) were not present within this specimen, which is consistent with the racemose cyst variant.

DISCUSSION
There is abundant medical literature on the treatment of NCC in general, however, the racemose form of NCC

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**Figure 1:** Pre-operative MRI brain with and without contrast was obtained prior to intervention, showing a large left multi-lobulated cystic lesion, causing significant mass effect and midline shift. The cyst contents appear isointense to CSF on MRI imaging. No evidence of enhancement was evident on contrasted imaging. Sequences are as follows: (a) Axial T2 sequence, (b) Axial FLAIR, (c) Axial with contrast, and (d) Coronal with contrast

**Figure 2:** Left craniotomy and sylvian fissure dissection is shown. (a) Evagination of the cyst wall with initial sylvian dissection. (b) Gentle traction is applied to the cyst to aid in removal. (c) The cyst ruptures during the process of removal. Copious irrigation was performed to clear cyst contents and the patient was given another dose of dexamethasone. (d) Gross specimen of multi-cystic racemose neurocysticercosis after en bloc resection
is less commonly encountered, thus limited literature and no evidence-based guidelines exist discussing the appropriate treatment of this enigmatic and deadly disease.\cite{3,4,17,25} In the past few decades, management of patients with NCC using anti-parasitic drugs, improved anti-inflammatory treatments, and minimally invasive neurosurgery has improved the prognosis of those infected with *T. solium*.\cite{9,12} Prognosis, however, varies in relation to the location and burden of parasites, with subarachnoid and intra-ventricular NCC exhibiting higher morbidity and mortality.\cite{12}

**Diagnosis**

Diagnosis of extraparenchymal neurocysticercosis can be difficult, given that histological confirmation is often not possible without surgical intervention, and is typically based on imaging and serologic findings.\cite{10} Recently, consensus diagnostic criteria was established and validated to help confirm the diagnosis of extraparenchymal NCC. The criteria are as follows, with one criteria being sufficient for diagnosis: an extraparenchymal cyst with pathologic diagnosis; at least one extraparenchymal cyst on MRI with special sequences exhibiting a scolex; or at least one cyst on MRI without a scolex exhibiting at least two of the following: hydrocephalus, inflammatory CSF pattern, positive immunologic testing, or presence of calcifications.\cite{5} Sensitivity of diagnosis using these criteria, specifically for the extraparenchymal form, still appears relatively low at 65.9%.\cite{5} Imaging findings of the subarachnoid and ventricular variants are often subtle, given that cysts have a similar density to CSF and may not be easily visualized on CT or MRI.\cite{10,15} This is not typically an issue with the racemose variant, as imaging typically shows a large multi-lobulated cystic lesion exerting mass effect and midline shift.\cite{19} CT imaging may detect calcifications if the cysts are older and dormant, but is of little use to differentiate RNCC from other possible intracranial lesions.\cite{18,19} MR imaging remains the modality of choice, as T2, FLAIR, and FIESTA sequences can more easily distinguish cysts within the subarachnoid space or ventricles.\cite{6,18,19} Cyst walls may show contrast enhancement, but this is dependent on the amount of inflammation associated with the lesion, and may not enhance at all in dormant lesions with little associated inflammation.\cite{18,19}

Serological studies detect antibodies to the *T. solium* within serum or CSF. The best serological test for NCC is the enzyme-linked immunoelectrotransfer blot (EITB) assay to detect serum antibodies, with sensitivity reported around 98% for patients with two or more live parasites.\cite{12} Sensitivity, however, is lower in patients with one intracranial cysticercus (50–60%), thus a negative test cannot preclude diagnosis.\cite{12} Enzyme-linked immunosorbent assay (ELISA) is also an option for testing both CSF and serum, and has a high sensitivity (89%) and specificity (93%) in CSF, similar to EITB.\cite{12} Both studies may falsely be negative in patients with few cysts or calcified (dormant) lesions, but this has not been validated with RNCC specifically.\cite{12} Cisternal forms, such as the racemose variant, are more easily detected in CSF than the typical parenchymal form, given their location and association with CSF spaces.\cite{12,21}

**Medical therapy**

While guidelines exist regarding the management of NCC in general, there is no consensus on the management of RNCC regarding both surgical and medical means.\cite{10,22,24} Typical medical treatment involves management of symptoms and associated disease sequelae, such as seizures and edema, as well as anti-parasitic therapy. First line anti-epileptic drugs should be used for seizures during periods of active infection and kept for at least 2 years in the case of seizures caused by inactive or calcified lesions.\cite{24} Peri-lesional
The most common anti-parasitic drugs include Dexamethasone at 0.1 mg/kg/day. The parasiticidal efficacy of albendazole and praziquantel also showed significant. Others, however, suggest treatment with albendazole alone and no surgical intervention for cyst involution treating racemose NCC with extended albendazole treatment (30 mg/kg/day) may not be curative even with several cycles of treatment. Albendazole mono-therapy was chosen, as it is the affected area also proved to be essential in safe resection. A generous craniotomy to expose the cyst with a Touhey needle, which proved necessary to provide the needed decompression to safely attempt sylvian dissection. A generous craniotomy to expose the affected area also proved to be essential in safe resection. Albendazole mono-therapy was chosen, as it is not a concern, due to a lack of daughter parasites within the cyst. This is in contrast to hydatid cysts, which are filled with daughter organisms that can seed other locations. In our patient, we chose open surgical removal given the low chance of success with medical therapy alone, the accessible location of the lesion, and extreme amount of mass effect and midline shift caused by it. Dexamethasone therapy was initiated given evidence of peri-lesional edema on MRI and associated mass effect. Treatment with pre and postoperative steroids, as well as intra-operative irrigation proved sufficient in avoiding the inflammatory sequelae of cyst rupture. It is likely that rupture was unavoidable in this particular case and may have simply been due to the initial decompression of the cyst with a Touhey needle, which proved necessary to provide the needed decompression to safely attempt sylvian dissection. A generous craniotomy to expose the affected area also proved to be essential in safe resection. Albendazole mono-therapy was chosen, as it is a common treatment for NCC, although recent studies have shown efficacy with albendazole and praziquantel in combination, and may be also considered as an option.

**CONCLUSIONS**

Racemose NCC remains a less common form of NCC and is an especially difficult disease to treat. No consensus exists on the proper treatment protocols for this particular variety of NCC. Further study is needed to define a consensus standard of care for patients with this entity. We present a case of RNCC in a non-endemic
region treated successfully with both open surgical and medical therapy, as well as management of intra-operative cyst rupture.

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**Conflicts of interest**
There are no conflicts of interest.

**REFERENCES**

1. Agapejev S. Neurocysticercosis: The enigmatic disease. Cent Nerv Syst Agents Med Chem 2011;11:261-84.
2. Anqi X, Jiahe X, Xiaoke Z, Chao Y. The surgical value of Neurocysticercosis: Analyzing 10 patients in 5 years. Turk Neurosurg 2013 [Epub ahead of print].
3. Baird RA, Wiebe S, Zunt JR, Halperin JJ, Gronseth G, Roos KL. Evidence-based guideline: Treatment of parenchymal neurocysticercosis: Report of the Guideline Development Subcommittee of the American Academy of Neurology. Neurology 2013;80:1424-9.
4. Bansal R, Gupta M, Bharat V, Sood N, Agarwal M. Racemose variant of neurocysticercosis: A case report. J Parasit Dis 2016;40:546-9.
5. Carpio A, Fleury A, Romo ML, Abraham R, Fandiño J, Durán JC, et al. New diagnostic criteria for neurocysticercosis: Reliability and validity. Ann Neurol 2016;80:434-42.
6. Carrillo Mezo R, Lara García J, Arroyo M, Fleury A. Relevance of 3D magnetic resonance imaging sequences in diagnosing basal subarachnoid neurocysticercosis. Acta Trop 2015;152:60-5.
7. Colli BO, Carlotti CG, Assirati JA, Machado HR, Valença M, Amato MCM. Surgical treatment of cerebral cysticercosis: Long-term results and prognostic factors. Neurosurg Focus 2002;12:E3.
8. Couldwell WT, Zee CS, Apuzzo ML. Definition of the role of contemporary surgical management in cisternal and parenchymatous cysticercosis cerebri. Neurosurgery 1991;28:231-7.
9. Fleury A, Carrillo-Mezo R, Flisser A, Sciutto E, Coronato T. Subarachnoid basal neurocysticercosis: A focus on the most severe form of the disease. Expert Rev Anti Infect Ther 2011;9:123-33.
10. Garcia HH, Evans CAW, Nash TE, Takayanagui OM, White AC, Botero D, et al. Current consensus guidelines for treatment of neurocysticercosis. Clin Microbiol Rev 2002;15:747-56.
11. Garcia HH, Lescano AG, Gonzales I, Bustos JA, Pretell EJ, Horton J, et al. Cysticidal Efficacy of Combined Treatment With Praziquantel and Albendazole for Parenchymal Brain Cysticercosis. Clin Infect Dis 2016;62:1275-9.
12. García HH, Nash TE, Del Brutto OH. Clinical symptoms, diagnosis, and treatment of neurocysticercosis. Lancet Neurol 2014;13:1202-15.
13. Góngora-Rivera F, Soto-Hernández JL, González Esquivel D, Cook HJ, Márquez-Caraveo C, Hernández Dávila R, et al. Albendazole trial at 15 or 30 mg/kg/day for subarachnoid and intraventricular cysticercosis. Neurology 2006;66:436-8.
14. Gonzales I, Rivera JT, Garcia HH, Cysticercosis Working Group in Peru. Pathogenesis of Taenia solium taeniasis and cysticercosis. Parasite Immunol 2016;38:136-46.
15. Hawk MW, Shahlaie K, Kim KD, Theis JH. Neurocysticercosis: A review. Surg Neurol 2005;63:123-32; discussion 132.
16. Jiménez-Vázquez OH, Nagore N. Endoscopic evidence of ventricular and cisternal inflammatory changes after intraoperative cysticercal rupture during endoscopic third-ventriculostomy removal. Br J Neurosurg 2013;27:137-8.
17. Krupa K, Krupa K, Pisculli ML, Athas DM, Farrell CJ. Racemose neurocysticercosis. Surg Neurol Int 2016;7:12.
18. Lerner A, Shiroishi MS, Zee CS, Law M, Go JL. Imaging of neurocysticercosis. Neuroimaging Clin N Am 2012;22:659-76.
19. Machado DC, Camilo GB, Alves UD, de Oliveira CE, de Oliveira RV, Lopes AJ. Imaging aspects of the racemose neurocysticercosis. Arch Med Sci 2015;11:1336-60.
20. Mahale RR, Mehta A, Rangaswamy T. Cysticercosis: A comprehensive review. J Neurol Sci 2015;364:11-21.
21. Michelet L, Fleury A, Sciutto E, Kendijo E, Fragoso G, Paris L, et al. Human neurocysticercosis: Comparison of different diagnostic tests using cerebrospinal fluid. J Clin Microbiol 2011;49:195-200.
22. Prazlov JV, Madrazo I, Avelar F, López-Félix B, Díaz G, Grijalva I. Medical treatment for neurocysticercosis characterized by giant subarachnoid cysts. N Engl J Med 2001;345:879-85.
23. Rajshhekar V. Surgical management of neurocysticercosis. Int J Surg (Lond Engl) 2010;8:100-104.
24. Rangel-Castilla L, Serpa JA, Graviss EA, Diaz-Marchan P, White AC. Contemporary neurosurgical approaches to neurocysticercosis. Am J Trop Med Hyg 2009;80:373-8.
25. Sharma S, Modi M, Lal V, Prabhakar S, Bhardwaj A, Sehgal R. Reversible dementia as a presenting manifestation of racemose neurocysticercosis. Ann Indian Acad Neurol 2013;16:88-90.
26. Sinha S, Sharma BS. Neurocysticercosis: A review of current status and management. J Clin Neurosci 2009;16:867-76.
27. Takayanagui OM, Odashima NS. Clinical aspects of neurocysticercosis. Parasitol Int 2006;55:S111-5.
28. Tuzun Y, Kadioglu HH, Izci Y, Suma S, Keles M, Aydin IH. The clinical, radiological and surgical aspects of cerebral hydatid cysts in children. Pediatr Neurosurg 2004;40:155-60.
29. Umredkar A, Singla N, Mohindra S, Bal A, Gupta SK. Giant intraparenchymal neurocysticercosis: Report of surgical aspects two cases. Neurol India 2009;57:800-2.