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Intraventricular Neurocysticercosis Presenting as Rapidly Progressive Dementia

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Abstract

We present a 38-year-old Guatemalan patient who presented to our hospital with new onset dementia and confusion of 2 months duration. A diagnosis of neurocysticercosis was made based on neuroimaging, patient's demographic information, and ELISA results. While dementia has been reported in 12.5% of cases with neurocysticercosis, rapidly progressive dementia as the sole presenting symptom is rare. Our patient was treated with Dexamethasone, Albendazole, and External Ventricular Drain (EVD) followed by Ventriculoperitoneal (VP) shunt to reduce his Intracranial Pressure (ICP). Eventually, resection of the cyst was performed. His mental status and cognition improved dramatically, and he was discharged from our hospital on a 2-week course of Albendazole. Neurocysticercosis patients with dementia and increased ICP should receive early aggressive medical and surgical intervention, which has been to improve outcome.

Keywords: Dementia; External ventricular drain; Hydrocephalus; Neurocysticercosis; Parasite infection

Introduction

Neurocysticercosis (NCC) is a zoonosis caused by the encysted larval stage of the tapeworm *Taenia Solium*. Incidence of infection in the United States is typically associated with travelers coming from endemic countries in Latin America, Africa, and Asia. Humans become infected through oral-fecal transmission, usually through consumption of undercooked or poorly handled food contaminated with the eggs of the parasite. After the parasites hatch and penetrate through the intestinal mucosa of the human gut, they gain access to the bloodstream and travel to different organs. Collectively, the clinical manifestations are known as cysticercosis, with the term "neurocysticercosis" referring to those pertaining to the central nervous system.

Clinical manifestations of NCC depends on the location of infection in the brain, for example, invasion of brain parenchyma causing seizures versus cyst formation outside of the parenchyma causing hydrocephalus [1]. We report a case of rapid cognitive decline as the sole presentation of NCC due to an intraventricular cysticercosis cyst.

Materials and Methods

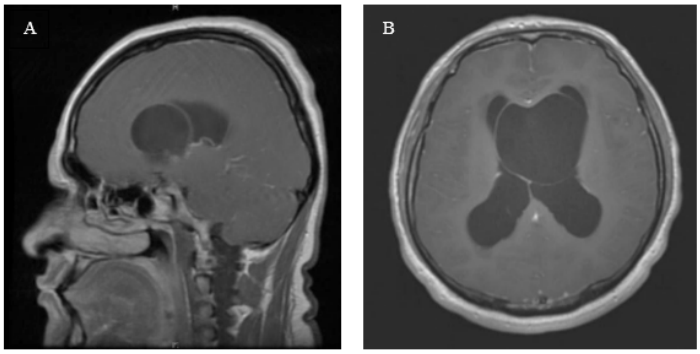
Case report with literature review were performed

Results

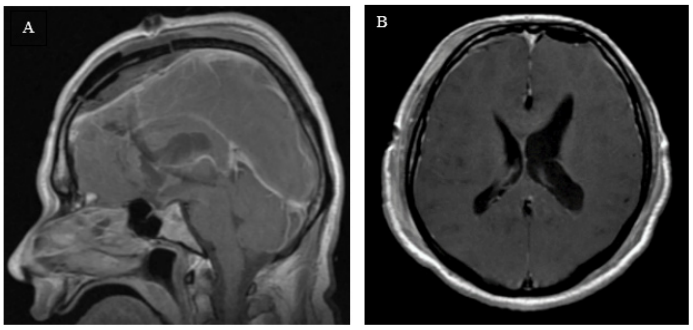
A 38-year old Guatemalan man presented to the Emergency Department on December 27, 2015 with 2 months of progressive memory loss, confusion, and wandering. History was obtained from his brother and cousin. They denied any history of seizures, central nervous system infections, or previous symptoms of confusion and memory loss. His initial neurological examination showed a Mini Mental Status Examination score of 16/30, and patient was unable to complex commands. CT Head without contrast showed a cystic mass occupying bilateral frontal horns of the lateral ventricles. Serum Enzyme-Linked Immunosorbent Assay (ELISA) to detect antibodies to *Taenia Solium* was positive. Examination of Cerebrospinal Fluid (CSF) was unremarkable with normal white blood cells, glucose, and protein. CSF Enzyme-Linked Immunoelectrotransfer Blot (EITB) was not performed.

MRI Brain showed a cystic structure occupying bilateral frontal horns of the lateral ventricles, with severe hydrocephalus leading to third ventricle obstruction and tentorial herniation as well as multiple calcified lesions, suggestive of NCC (Figures 1a and 2b). Administration of Gadolinium revealed enhancement of a thin wall circumscribing the lesion.

The patient was initially treated with Dexamethasone 2mg intravenously every 6 hours for 10 days. A right occipital External Ventricular Drainage (EVD) was subsequently placed resulting in a marked decrease in the size of the cyst and resolution of the 3rd ventricular obstruction. One week later he underwent resection of the cyst using an interhemispheric approach. A right frontal EVD and right occipital Ventriculoperitoneal Shunt (VPS) were placed during the procedure to relieve increasing intracranial pressures. This was followed by Albendazole 400mg orally twice daily for four weeks. Three cysts with multiple nodules which were removed during the procedure were submitted to the neuropathologist. Gross examination of the cysts revealed tan yellow solid masses consistent with the pathology of a cyst. Histopathological examination of the cyst reveals the body of the embryo of *Taenia Solium* and parts of the cyst wall (Figures 2A and 2B). During his hospital course, patient's orientation and judgment improved and he was able to follow complex two-step commands. He was discharged on a 2-week course of Albendazole 400mg orally twice daily.



Figures 1: A-B: Pre surgery - MRI Brain with and without contrast - Post contrast, T1 axial and sagittal sections.



Figures 2: A-B: Post-surgery - MRI Brain with and without contrast - Post contrast, T1 axial and sagittal sections.

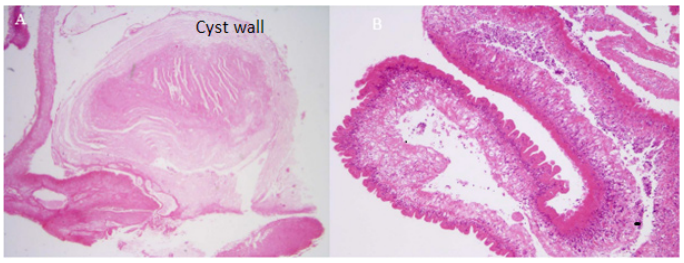


Figure 3: A-B: Microscopic H&E staining of a part of the cyst wall. (A) shows the body of the embryo along with part of the cyst wall, scolex was not obtained in the image (B) shows a portion of the cyst wall.

Discussion

We report an unusual presentation of NCC with rapidly progressive cognitive decline due to CSF obstruction and hydrocephalus that was reversible with surgical resection and treatment. Dementia has been reported in 12.5% of cases with NCC [2] and is thought to be due to multiple parenchymal parasitic lesions. Rapidly progressive dementia as the only presentation of NCC is rare and caused by increased ICP due to an obstructive cyst impeding CSF drainage. It is important to recognize this presentation since the dementia may be reversible [3]. Hydrocephalus and increased ICP due to obstruction of CSF flow has been reported in 30% of patients with NCC [4]. Dementia due to obstructive hydrocephalus has an excellent outcome when detected and treated early, unlike dementia due to multiple parenchymal parasitic lesions.

Cysticercosis is the collective term of clinical manifestations caused by the encysted larval stage of the tapeworm *Taenia Solium*. The parasite can affect any organ of the body, but the brain is the most common site of infection in the central nervous system with its set of symptoms termed NCC [5]. NCC may manifest differently depending upon the location of the infection in the brain. Parenchymal NCC is most located in the cerebral hemispheres, usually at the gray-white matter junction. Although rare, there are a few case reports of NCC with lesions in the cerebellum [6]. Extraparenchymal NCC can be in the ventricles, the cisternal or subarachnoid spaces, spinal cord, and ocular bulb. Subarachnoid locations are less common and generally occur in the basal cisterns or Sylvian fissure [7]. Racemose NCC is a severe variant form of extraparenchymal NCC, with the name descriptive of its macroscopic appearance as a "cluster of grapes", is associated with hydrocephalus, and has a high mortality rate (50%) [8].

The onset of symptomatic NCC has been estimated to peak at three to five years after infection, but may can be delayed for more than 30 years. Clinical manifestations of parenchymal NCC depend on the location and number of parasitic lesions and the associated inflammatory response. The most common presentation is seizures [7]. On the other hand, racemose NCC, due to its extra-axial location, typically presents with symptoms of raised intracranial pressure and meningitis. The subarachnoid space cysts

may give rise to focal mass effect or to an inflammatory response that presents clinically as a basilar meningitis. Non-communicating hydrocephalus and signs of elevated intracranial pressure, such as nausea, vomiting, headache, and papilledema, may follow. Cranial neuropathy or vasculitis may result from the inflammatory changes, leading to small- or large-vessel infarcts [8-10].

Cognitive impairment has been reported in two-thirds of patients with NCC [2]. Dementia as a sole presentation is very rare, especially in racemose NCC. The mechanisms proposed for cognitive decline in NCC include raised intracranial pressure, number and location of NCC lesions, the different phases of evolution of the parasite, inflammatory cytokines, and the host's immune response to the parasite [2]. A study by C.L Rodrigues and his group showed that NCC leads to a spectrum of cognitive abnormalities, ranging from impairment in a single cognitive domain to dementia [11]. This study also showed that dementia only presented in patients with active NCC infection. Patients with calcified, or inactive, NCC lesions had only cognitive impairment without fulfilling the rest of the diagnostic criteria for dementia [11]. The prognosis of cognitive impairment in NCC depends upon the underlying mechanism and administered treatment.

The diagnosis of NCC is largely based on clinical presentation and radiographic imaging. Serologic tests and invasive procedures such as a brain biopsy may be helpful to confirm the diagnosis, especially when neuroimaging is non-diagnostic. Del Brutto et al. proposed a set of criteria to diagnose NCC in 2011 including the use of both MRI and CT [12]. Both imaging modalities should be performed, as CT head identifies calcified lesions effectively and MRI brain better detects extra-axial cysts [13]. In addition to imaging, several serologic tests have been developed for both serum and CSF. Some assays, such as ELISA, can detect anti-cysticercal antibodies, while others can identify cysticercal antigens. The test of choice for antibody detection is the EITB, which can be performed on serum or CSF [14]. Antibodies may be detected several months after the active infection, while antigen detection tends to have higher sensitivity and specificity for active cysts and those in extraparenchymal locations [14,15].

Management of NCC consists of several important components, such as anti-inflammatory, anti-epileptic, anti-parasitic drugs and neurosurgical intervention [16]. Our patient presented with rapid cognitive decline secondary to hydrocephalus and initial treatment was aimed at reducing ICP by diverting CSF flow through a VP shunt [17]. It is not recommended to administer albendazole prior to either endoscopic or open dissection of the cyst, as the drug can cause inflammatory changes in the brain thus complicating intraoperative and postoperative outcomes [18].

Corticosteroids, such as prednisone 1mg/kg/day or dexamethasone 0.1mg/kg/day, for five to ten days and tapered, are used prior to cyst resection to reduce inflammation [19]. After resection, anti-parasitic drugs, such as albendazole, may be initiated at 15mg/kg/day or else 800mg/day divided in two doses and continued depending upon the type and number of lesions [20]. Anti-epileptic medications may be initiated in patients who are at high risk of seizure and should be continued six to twelve months after the infection is radiographically resolved, although a recent Cochrane review showed no significant difference in NCC patients treated for six to twelve months as opposed to a longer time period of 24 months [21,22].

Conclusion

Although dementia has been reported to accompany 12.5% of cases with NCC, rapidly progressive dementia as a sole presentation of NCC is rare. Rapid treatment of raised ICP and administration of corticosteroids and albendazole can lead to marked improvement of cognitive symptoms in NCC patients with hydrocephalus, as compared to multiple parenchymal parasitic lesions [3].

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