Case Report: An Unusual Presentation of Neurocysticercosis: A Space-Occupying Lesion in the Fourth Ventricle Associated with Progressive Cognitive Decline

Carolin Kurz,* Veronika Schmidt, Holger Poppert, Patricia Wilkins, John Noh, Sven Poppert, Jürgen Schlegel, Claire Delbridge, Clarissa Prazeres da Costa, and Andrea S. Winkler

Department of Neurology, Klinikum rechts der Isar, Technical University Munich, Germany; Division of Parasitic Diseases and Malaria, Centers for Disease Control and Prevention, Atlanta, Georgia; University Medical Center, Hamburg-Eppendorf, Germany; Division of Neuropathology, Institute of Pathology, Technical University Munich, Germany; Institute of Medical Microbiology, Immunology and Hygiene, Klinikum rechts der Isar, Technical University Munich, Germany

Abstract. We communicate a case of a middle-aged Brazilian patient with an unusual presentation of fourth ventricular neurocysticercosis: occurrence of two intraventricular cysts at different locations in the brain within 2 years and cognitive decline as the only neurological symptom. Neurocysticercosis was confirmed by magnetic resonance imaging, serology, histology, and genetic analysis. Neurocysticercosis should be considered as a differential diagnosis in cases with atypical neurologic or psychiatric symptoms, atypical neuroimaging and travel history. Especially, fourth ventricular cysts carry the risk of obstructive hydrocephalus and brainstem compression and therefore should be extirpated completely. If complete removal of the cystic structures cannot be proven in cases with surgically treated neurocysticercosis, anthelminthic therapy and thorough follow-up examinations should be conducted.

INTRODUCTION

Taeniasis and cysticercosis are diseases caused by two different stages of the same parasite, the pork tapeworm *Taenia solium*; taeniasis denotes intestinal infection with the adult tapeworm, while cysticercosis represents tissue infection with the larval stage.1 Neurocysticercosis (NCC) implies cerebral and/or spinal infection with larvae of *T. solium* and is the most common helminthic infection of the central nervous system (CNS).2–4 NCC is endemic in many low-income countries and increasingly diagnosed in high-income countries due to travel and migration.5 Cysticerci pass through various stages, and in their mature form, they can persist for many years in the host’s body.3 If cysticerci degenerate, an inflammatory reaction may cause symptoms/signs varying from headaches and drowsiness to seizures, obstructive hydrocephalus and stroke. The extent of inflammation depends on the number, localization and size of the cysts as well as on the immunological response of the host.2,5–7 In their late stage, cysticerci calcify and may or may not leave a residual nodule.2,4 Only symptomatic NCC requires treatment with anthelmintic drugs, steroids, and antiepileptic medication (if epileptic seizures are present) or surgical removal of cysts.2–4,8,9

*Taenia solium* cysticerci can settle in different regions of the human brain: intraparenchymal NCC (60–90% cases) may cause focal neurological signs, mostly headache and epileptic seizures; extraparenchymal NCC, located in the ventricular system or subarachnoid space, has a poorer prognosis, because this form of NCC carries the risk of severe complications such as hydrocephalus or vasculitis and is more difficult to treat than the intraparenchymal form.2–5,9

The literature concerning the management of intraventricular NCC is ambiguous and a clear stratification into the different forms of ventricular cysts (lateral and third/fourth ventricular cysts) or into the presence or absence of concomitant parenchymal cysts is missing. In general, there are three treatment options: 1) medical therapy, 2) surgical removal of the cyst and 3) ventriculoperitoneal (VP) shunt placement.10–12

The role of anthelminthic therapy in intraventricular NCC remains uncertain: intraventricular NCC may respond favorably to albendazole, but both failures and successes with praziquantel have been reported.13 Apart from that, albendazole is preferred over praziquantel because of its better penetration into the CNS, greater cysticidal effect, and less interaction with other drugs such as steroids.9,10 Further beneficial effects of anthelmintic drugs are the destruction of additional cysts, if present.13,14 Corticosteroids are used to relieve inflammatory symptoms/signs caused by larval death (headache, nausea, vomiting and seizures), that usually occur 2–5 days after initiation of anthelmintic therapy.10,13

Surgical removal of cysts is required in complicated cases with acute obstructive hydrocephalus.2–5 Especially, fourth ventricular cysts should undergo extirpation, because this form may cause brainstem compression even after insertion of a VP shunt.13 Furthermore, a VP shunt is usually required in patients with chronic hydrocephalus and chronic increased intracranial pressure.13

In a recently published meta-analysis about the treatment of intraventricular NCC, the authors proposed medical treatment in cases of intraventricular NCC without hydrocephalus or in cases with surgically non-accessible cysts and surgical treatment in cases with hydrocephalus. They suggested postsurgical medical therapy (anthelmintic medication and steroids) in cases with incomplete resection or spilling and/or the presence of further cysts.14

CASE PRESENTATION

A middle-aged patient with migration background (Brazil) presented to our department, because he had been unable to work due to poor concentration for 2 months. Remarkable in the patient’s past medical history was an episode with severe headache, confusion, somnolence, and vomiting and one generalized epileptic seizure in June 2011; he was treated in a municipal hospital. Then brain imaging had revealed obstructive hydrocephalus caused by a mass in the third ventricle (Figure 1A and B). The mass was removed by endoscopic...
ventriculostomy, but pathological findings were inconclusive and a hemangioblastoma was considered the most likely diagnosis. After that episode the patient had neither suffered from epileptic seizures nor from symptoms indicating obstructive hydrocephalus. Follow-up magnetic resonance imaging (MRI) scans were not available.

**Diagnostic findings.** On admission to our hospital in February 2013, neurological examination was unremarkable despite that the patient seemed absentminded and distracted. On neuropsychological testing, the patient performed below age, gender, and education reference in executive function, learning tasks, alertness, celerity and working memory. A standardized language test was not applied because of the language barrier. We interpreted these findings as a moderate cognitive decline. Electroencephalography recorded intermittent generalized slowing of 4–5 Hz without epileptic activity. MRI identified a small contrast-enhancing mass at the bottom of the fourth ventricle (10 × 11 × 8 mm) adjacent to a cyst (23 × 17 × 11 mm). The lateral ventricles as well as the fourth ventricle were slightly dilated (Figure 1C and D). We did not detect additional cerebral or spinal cysts.

Because of uncertain diagnosis, the patient underwent ventriculostomy of the fourth ventricle, and histologic workup revealed intraventricular NCC (Figure 2A–D). A papillary cystic wall with necrotic areas was identified that was surrounded by a chronic inflammatory reaction with enclosed calcified corpuscles (Figure 2A–D). The cyst wall was lined by an eosinophilic layer with hair-like protrusions (microtrichia) (Figure 2A and B; hematoxylin and eosin staining), followed by a reticular inner layer. Within this, periodic acid–Schiff reaction staining revealed hyphae-like structures resembling excretory ducts. A scolex (head of the larva) could not be identified. Taken together, we interpreted these structures as a racemose cyst with surrounding inflammatory reaction.

Sequence analysis of mitochondrial Cox1 gene was performed and showed 100% similarity to the reference sequence of *T. solium* (Cox1 gene, GenBank no. AB0066492.1); genotypically the sequence exhibited 100% consensus with the “Brazilian type” of *T. solium.*

Cerebrospinal fluid (CSF) was performed after surgery and showed an elevated cell count (39 cells/μL), elevated protein concentration (761 mg/L), and eosinophilia. White blood count was 10,700/μL with 2% eosinophils. Immunoblot (LD Bio® Diagnostics, Lyon, France) for antibodies against *T. solium* was positive in serum and CSF. Lentin-lectin-glycoprotein enzyme-linked immuno-electrotransfer blot and recombinant rT24-immunoblot confirmed the diagnosis of NCC in serum and CSF. In addition, an antigen enzyme-linked immunosorbent assay (apDia®, Turnhout, Belgium) revealed a positive result in serum but not in CSF. There were no antibodies...
against the adult stage of the tapeworm (taeniasis; RES33-immunoblot) verifiable in the patient’s serum. Serological tests for echinococcosis, amebiasis, and other viral or bacterial infections including HIV and hepatitis were negative just as microscopic stool examination for Taenia eggs.

Taken all serological findings together, we interpreted the results as an infection of the CNS with the cystic/larval stage of T. solium. Together with the negative microscopic stool examination for Taenia eggs and no proof of an intestinal infection, we considered the patient as not contagious at the time of diagnosis.

Outcome. Since we had no evidence for other or residual cerebral and spinal lesions on postsurgery MRI or the presence of epileptic seizures and since the cyst had been surgically removed, we refrained from antiparasitic treatment. The patient was discharged to a rehabilitation center with an emphasis on cognitive training and reported improved cognition afterward. Unfortunately, the patient has been lost to follow-up, and a reexamination including CSF analysis, neuropsychological testing and MRI could not be performed.

DISCUSSION

We present a patient with an unusual cause of cognitive impairment. Patients with both intra- or extraparenchymal NCC can present with psychiatric manifestations such as cognitive decline and depression. The literature regarding the frequency of cognitive impairment in NCC is inconsistent and the relationship between cognitive decline and parasite-specific features remains unclear: mild forms of cognitive decline have been described in 45% and 71.9% of patients with NCC, whereas dementia or severe cognitive deterioration were diagnosed in 12.5% and 15.1% of affected individuals. Others reported cognitive impairment to a lesser degree (17% and 12.5%). Although in two studies cognitive decline did not correlate with parasite-specific features, one study found evidence that cognitive decline may arise from an interaction between number, localization and local inflammation of cystic lesions disrupting frontal-parietal-temporal networks.

In this patient, there was a striking discrepancy between the absence of parenchymal lesions or obstructive hydrocephalus and the patient’s cognitive impairment. We assumed a chronic inflammatory process, represented by the increased cell count and elevated protein concentration in the CSF, to be the most likely explanation for the patient’s cognitive impairment.

Another very unusual feature was the occurrence of two intraventricular cysts at two different locations without a clear source of reinfection. We suspect that the cyst was incompletely removed by the first ventriculostomy in 2011 and has resettled in the fourth ventricle. In the literature, one similar case with a migrating intraventricular cyst after surgical evacuation has been described. If complete removal of the cystic structures cannot be proven in cases with surgically treated NCC, anthelminthic therapy and follow-up examinations including brain imaging have been recommended.

In our case, genetic testing showed that the parasite was most probably derived from Brazil, so either the patient got infected decades ago while still living in the endemic country Brazil (he moved from Brazil to Europe in the late 1980s) or during his infrequent later visits (last in 2005). Therefore, the performance of genetic sequencing of the parasite added
useful information complementing histology, as it not only confirmed the suspected diagnosis but also indicated the region where the infection was acquired.

CONCLUSIONS

NCC should be kept in mind in patients with atypical neurological and/or psychiatric symptoms, atypical neuroimaging and with recent or past travel history. Especially, fourth ventricular cysts carry the risk of obstructive hydrocephalus and brainstem compression and should undergo extirpation. We recommend thorough morphologic and histologic investigations of surgical specimen for completeness. If complete removal of the cystic structures cannot be proven in cases with surgically treated NCC, anthelminthic therapy and follow-up examinations including brain imaging have to be considered. In summary, surgical and medical management of intraventricular NCC remains nonstandard and varies among institutions because of the lack of systematically conducted trials.

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Authors’ addresses: Carolin Kurz, Veronika Schmidt, Holger Poppert, and Andrea S. Winkler, Department of Neurology, Klinikum rechts der Isar, Technical University Munich, Germany, E-mails: kurz@lrz.tum.de, veronika.schmidt@tum.de, poppert@neurovasc.de, and andrea.winkler@tum.de. Patricia Wilkins and John Noh, Division of Parastic Diseases and Malaria, Center for Disease Control and Prevention, Atlanta, E-mails: pwillkins@cdc.gov and jnnoh@cdc.gov. Sven Poppert, University Medical Center, Hamburg-Eppendorf, Germany, E-mail: sven@poppert.eu. Jürgen Schlegel, Claire Delbridge, Division of Neuropathology, Institute of Pathology, Technical University Munich, Munich, Germany, E-mails: juergen.schlegel@lrz.tu-muenchen.de, c.delbridge@tum.de. Clarissa Prazeres da Costa, Institute of Medical Microbiology, Immunology and Hygiene, Klinikum rechts der Isar, Technical University Munich, Germany, E-mail: clarissa.daCosta@tum.de.

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