Case Report

Fourth ventricle neurocysticercosis: A case report

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Received: 01 July 18  Accepted: 03 September 18  Published: 03 October 18

Abstract

Background: Neurocysticercosis (NCC) is the most common helminthic disease of the nervous system in humans and it is caused by the larvae of the pork tapeworm, Taenia solium. We present a case of microsurgical removal of a fourth ventricle NCC cyst combined with an endoscopic third ventriculostomy (ETV) to treat hydrocephalus.

Case Description: A 36-year-old woman presented to the emergency room with headache and decreased visual acuity over the last 4 months. A brain magnetic resonance imaging showed obstructive hydrocephalus apparently correlated to a mobile, cystic lesion of the fourth ventricle. In the same operative time, an ETV and a suboccipital craniotomy were performed in order to remove the lesion and to treat the hydrocephalus. The cyst was completely removed and pathologically identified as a T. solium cyst. The early postoperative course was uneventful and she was discharged asymptomatic and off anthelmintic medication. Five weeks later, the patient returned with hydrocephalus recurrence and was successfully retreated with an ETV. At 5-month follow-up, she remains asymptomatic and has no evidence of persistent disease or hydrocephalus recurrence.

Conclusion: Intraventricular neurocysticercosis is, typically, a surgical disease. For cysts located on the fourth ventricle, a suboccipital craniotomy and a telovelar approach remains a valid option. Cyst removal does not necessarily resolve the hydrocephalus problem. ETV offers an option to the classic shunt placement approach and was shown to be effective even on hydrocephalus recurrence.

Key Words: Fourth ventricle, intraventricular neurocysticercosis, ventriculostomy

INTRODUCTION

Neurocysticercosis (NCC) is the most common helminthic disease of the nervous system in humans and is caused by the larvae of the pork tapeworm Taenia solium. Two main forms of the disease are known: intraparenchymal and extraparenchymal. Extraparenchymal disease can be subdivided in intraventricular, subarachnoid and spinal forms. Herein, we present a case of microsurgical removal of a fourth ventricle neurocysticercosis cyst combined with endoscopic third ventriculostomy (ETV) to treat hydrocephalus and a successfully redone ETV for hydrocephalus recurrence.

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How to cite this article: Simão D, Teixeira JC, Campos AR, Coiteiro D, Santos MM. Fourth ventricle neurocysticercosis: A case report. Surg Neurol Int 2018;9:201.

http://surgicalneurologyint.com/Fourth-ventricle-neurocysticercosis-A-case-report/
CASE

A 36-year-old female was referred to our emergency room by her homeland local hospital in Cape Verde (West Africa), with complaints of headaches and diminished vision over the last 4 months. On neurological examination, she had impaired visual acuity (right eye: 6/10; left eye: 7/10) and bilateral papilledema. No other neurological signs were observed. The brain magnetic resonance imaging (MRI) revealed hydrocephalus apparently caused by a well-defined cystic lesion in the fourth ventricle. The lesion was hypointense on T1-weighted and T2-weighted MRI sequences and had a peripheral millimetric solid component hyperintense on both sequences [Figure 1]. Another MRI was done 8 hours later due to a temporary malfunction of the gadolinium injector. In this second exam, the lesion showed no contrast enhancement but had changed its location to the contralateral side within the fourth ventricle [Figure 2], revealing its mobile nature. The FLAIR sequences showed cerebellar edema surrounding the cyst.

A surgical plan was elaborated to solve both problems – the hydrocephalus and the intraventricular cyst – at the same surgical time. Initially, an uneventful ETV was performed and intraventricular CSF was collected for further laboratorial analyses. No external drainage was left in place. The patient was then placed in prone position and underwent a suboccipital median craniotomy. Using microsurgical technique, a telovelar approach to the fourth ventricle was performed. The cyst microdissection was particularly challenging due to its thin wall and partial adherence to the surrounding ependyma, which contributed to an intraventricular cyst rupture by the end of its total removal. Extensive irrigation of the ventricle space with Ringer’s solution was performed to avoid intraventricular dissemination.

The patient had an uneventful postoperative course. The CSF analysis showed no alterations. Postop MRI showed a total removal of the cyst lesion and reduction of the ventricle dimensions [Figure 3]. The pathology report confirmed the cyst to be a T. solium larva in its vesicular stage as had already been suspected [Figure 4]. It was decided not to initiate antiparasitic medication given the lack of evidence of a positive effect in intraventricular single neurocysticercosis cyst. The patient was discharged asymptomatic seven days later with a prescription to a progressive corticosteroids’ weaning of.

Five weeks after discharge, she presented back, complaining of headaches and worsening of vision again. A significant pseudomeningocele with no CSF leak was visible underneath the frontal surgical wound. A bilateral papilledema was evident in the ophthalmological examination. The MRI showed hydrocephalus recurrence with no evidence of CSF flow through the previous ventriculostomy stoma [Figure 5]. A second endoscopic approach was performed to collect CSF and to reopen the ventriculostomy stoma, using the same burr hole and parenchyma trajectory. Intraoperatively, we visualized a transparent CSF plenty of floating debris [Figure 6]. In the histopathological examination, these were found to
be fragments of brain parenchyma with granulation tissue foci, lymphoplasmacytic inflammatory infiltrates, with histiocytes, few neutrophils, and multiple calcifications. The CSF analysis showed no cells and normal glucose and proteins’ levels. The patient did well postoperatively and was discharged 5 days later with no neurological deficits.

She has been followed up for 5 months now, remaining asymptomatic, with clearly defined optic discs on fundoscopy and has fully recovered visual acuity. The 5-month postop MRI ruled out hydrocephalus or any recurrent lesion [Figure 7].

**DISCUSSION**

NCC is the most common helminthic disease in the central nervous system and the most frequent preventable cause of epilepsy in the developing world.\(^{[21,24,27]}\) It is caused by the larval form of the *T. solium*, commonly referred as the “pork tapeworm.” Human NCC most commonly occurs through the ingestion of food or water contaminated with *T. solium* eggs.\(^{[14]}\)

Depending on the affected compartment, NCC may be classified as intraparenchymal or extraparenchymal and the latter subdivided in intraventricular, subarachnoid and spinal forms. Extraparenchymal disease is known to be associated with more severe complications, such as hydrocephalus, arachnoiditis, and ventriculitis,\(^{[28]}\) resulting in a worse overall outcome.\(^{[8]}\) Intraventricular cysts are most found in the fourth ventricle (43%–70%) followed by the lateral (11%–43%) and third (1%–29%) ventricles with a minority in the aqueduct (7%–9%).\(^{[16]}\) Hydrocephalus is frequently present in intraventricular NCC cases due to mechanical obstruction of CSF flow or associated arachnoiditis.\(^{[9]}\)

Surgical treatment in extraparenchymal NCC is the standard of care to intraventricular cysts, hydrocephalus due to racemose cysts, or hydrocephalus due to ependymitis.\(^{[11]}\) In this case, the patient presented...
with symptoms of elevated intracranial pressure due to hydrocephalus as an apparent result of mechanical obstruction of the fourth ventricle by a single intraventricular cysticercus. However, in the second preoperative MRI it was found that the cysticercus was not in a fixed position within the fourth ventricle. The possibility of mobile cysticerci within the ventricular space had already been reported.[7,22,25] In addition, preoperative MRIs showed periventricular changes [Figure 1], which could mean some degree of ependymitis, a known factor for hydrocephalus.[9,22] Therefore, we assumed that the surgical excision of the cyst might not treat the hydrocephalus and decided to perform a ventriculostomy in addition to the lesion removal. We decided to perform it at first since any intraventricular hemorrhage during cyst’s removal could compromise the visualization through the endoscope. Shunt placement for hydrocephalus caused by NCC is associated with high failure rates and the need for frequent shunt revisions.[10,15] By opposition, CSF diversion using endoscopy can achieve success rates >90% in some groups.[13,17] To our best knowledge, this is the first case published in which an ETV was added to an open posterior fossa approach to remove a fourth ventricle cysticercus and to avoid the use of a shunt. Furthermore, this case proves that a first ventriculostomy failure may not preclude a second attempt.

Minimally invasive endoscopic removal of intraventricular cysts has emerged as the preferred surgical approach, especially if the cysts are in the lateral or third ventricles.[6,13] Endoscopic resection of fourth ventricle cysts has been also reported. Kaf et al.[17] were able to excise fourth ventricle cysts in 8 of 12 patients using the endoscope as Goel et al.[13] in 14 patients, without any periaqueductal injury. However, the endoscopic technique seems to be more challenging and requires special expertise particularly in cases with a thin aqueduct[6,4,15] or if the cyst is adherent to the ependymal lining.[15,22] Depending on the surgeon’s experience, a posterior fossa open approach has been shown to be a good option. Loyo et al.[18] described 27 patients whom underwent suboccipital craniectomy with good or excellent outcomes in 81.4%, but >90% of them needed a shunting procedure. In Apuzzo et al. series,[2] 17 patients were treated with a primary posterior fossa approach and 6 of them had evidence of ependymitis and needed a CSF shunt. Other case reports[10,15] also describe a good outcome with a microsurgical posterior fossa approach. Nevertheless, complications such as periventricular edema, residual fourth ventricle cyst, fourth ventricle entrapment, need for reoperation and hematomas have been reported[5,18,20] and should be considered.

Despite some referred negative effects of an intraoperative cyst rupture,[19] most papers did not report major complications, such as ventriculitis.[17,21] Likewise, in the current case, the CSF analysis from the sample collected on the second surgery did not show signs of ventriculitis. One could speculate that the inflammatory exudate visualized with the endoscope resulted from the previous cyst rupture and that this event may have contributed to the ventriculostomy ostium closure. There is no similar report previously published.

Medical treatment for NCC remains controversial. In active intraparenchymal disease, anthelmintic medication has shown to prevent epilepsy and reduce the number of viable lesions.[1,3,12] The recent guidelines by the Infectious Diseases Society of America (IDSA) and the American Society of Tropical Medicine and Hygiene (ASTMH)[26] recommend albendazole monotherapy (15 mg/kg/day divided into two daily doses) for 10–14 days in case of one or two active parenchymal cysticerci. When more than two viable lesions are present, a combined treatment with albendazole (15 mg/kg/day) and praziquantel (50 mg/kg/day) is preferred for the same period. However, anthelmintic medication has not shown major benefits for intraventricular NCC susceptible for surgical removal.[6,22,26] Some experts even recommend to not use them preoperatively, as it can result in disruption of parasite integrity and an inflammatory response that can prevent successful cyst removal.[26] Steroids, as prednisolone or dexamethasone, are normally used to control the inflammatory process following degradation of viable cysts and as adjuncts to cysticidal therapy.[14] In our case, at 5-month follow-up, the patient had no signs of disease recurrence and no anthelmintic medication had ever been used.

**CONCLUSION**

Treatment of intraventricular NCC involves cysts removal and treatment of associated hydrocephalus. In the presence of a removable single intraventricular cyst, anthelmintic medication is normally not necessary. Endoscopic techniques to treat hydrocephalus, such as ETV, should be preferred to shunt placement. We present a case in which combined microscopic and endoscopic approaches were performed for removal of a fourth ventricle cysticercus and treatment of the associated hydrocephalus. Our case is an example that failure at a first ETV does not make a second attempt futile and that cyst rupture does not necessarily lead to a worse outcome, although its role in hydrocephalus relapse is uncertain.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be
made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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