Management of Trapped Fourth Ventricle in Patient with Cerebral and Spinal Neurocysticercosis

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Citation

Abstract
Neurocysticercosis (NCC) is the most common parasitic infection of the CNS. Surgical intervention is often required to treat for hydrocephalus with neuroendoscopy emerging as a logical alternative to craniotomies. While cranial NCC cases have increased in the United States, spinal NCC incidences have remained relatively low. We report a case of trapped fourth ventricle and myelopathy from both concomitant cranial and spinal NCC. This patient exhibited a subacute thoracic myelopathy and acute deterioration from a trapped fourth ventricle (TFV) from aqueductal and fourth ventricular outflow occlusion. She underwent urgent posterior fossa exploration. A fourth ventricular lesion consistent with NCC was removed from the distal aqueduct and the aqueduct was opened via aqueductoplasty with a ventricular catheter and balloon dilatation. She improved following surgery without extraocular dysfunction. Intraoperative visualization and postoperative MRI flow-studies confirmed patency of the aqueduct and there was complete resolution of her fourth ventricle dilation. With antiparasitic and corticosteroid therapy, her myelopathy has slowly improved over 9 months of follow-up.

ABBREVIATIONS
CSF: cerebrospinal fluid
CT: computed tomography
MRI: magnetic resonance imaging
NCC: neurocysticercosis
TFV: trapped fourth ventricle

INTRODUCTION
Paranoli first described cysticercosis in the central nervous system (CNS) in 1550, but Taenia solium was not recognized as the cause until the late 19th century by Leuckart and Küchenmeister. Cysticercosis is the most common parasitic CNS infection in the world. Endemic in Mexico, Latin America, tropical Africa, India, and Southeast Asia, neurocysticercosis (NCC) has recently begun to become more prevalent in the southwestern United States. Approximately one-third of the world population lives in an area where T. solium is endemic, causing 50 million infections and 50,000 deaths annually. The disease begins first with taeniasis, which arises from the human ingestion of undercooked pork containing cysterceral larvae that then develop into the adult worm in the human small intestine. Eggs are excreted into human stool and ingested by pigs or humans. Cysticercosis is the stage in which the eggs hatch and the larvae penetrate the intestinal mucosa to enter the bloodstream and migrate to muscle, brain, or eyes. NCC disseminates through the CNS via small capillaries to enter the parenchyma or via the choroid plexus to enter the ventricles and subarachnoid space. Approximately 60 to 92% of NCC patients have brain parenchymal involvement, while up to 20% of patients have intraventricular NCC. The disease is often self-limited unless symptomatic hydrocephalus develops, necessitating surgical intervention. NCC may also result in epileptic seizures, meningitis, intracranial hypertension, and focal neurologic deficits. In endemic regions, NCC is the most common cause of seizures and hydrocephalus. Spinal NCC often presents with myelopathy and progressive weakness from compression of the spinal cord or cauda equina.

We report a case of TFV and myelopathy from NCC. This patient exhibited a subacute thoracic myelopathy and acute deterioration from a TFV from aqueductal occlusion. She underwent urgent posterior fossa exploration. A lesion consistent with NCC was removed from the distal aqueduct and the aqueduct was opened via aqueductoplasty with a ventricular catheter and balloon dilatation. She improved following surgery without extraocular dysfunction.
Intraoperative visualization and postoperative MRI flow-studies confirmed patency of the aqueduct and there was complete resolution of her fourth ventricle dilatation. With antiparasitic and corticosteroid therapy, her myelopathy has slowly improved over 9 months of follow-up.

CASE REPORT

History and Presentation: This 40-year old female initially presented to an outside hospital 5 years prior complaining of diffuse headache. She was found to have multiloculated cysts within the basal cisterns and obstructive hydrocephalus.

Figure 1

Figure 1. CT showed enlarged lateral ventricles (A), normal caliber of the 4 ventricle (B) and multiloculated cysts within the basal cisterns (B). The latter was better appreciated on this FLAIR MRI image (C). Sagittal MRI with gadolinium suggested patency of the aqueduct and no contrast-enhancing lesions were appreciated.

NCC was suspected. A ventriculoperitoneal shunt was placed at that time with resolution of her headaches. The records were unavailable and it was unclear if the patient completed the prescribed antihelminthic treatment.

During the two years prior to her most recent presentation, she had experienced progressive deterioration in her gait and relied on a walker for ambulation. She presented to our center complaining of three days of acute headache and one day of severe nausea and vomiting.

Neurological Examination: On examination, she was awake and alert but uncomfortable. Her cranial nerve examination was unremarkable and her upper extremities exhibited normal sensorimotor functioning and no pathological reflexes. She exhibited 4/5 motor power in her iliopsoas and quadriceps and 4-/5 power in dorsiflexion and plantar flexion. The Babinski reflex was present bilaterally, knee and ankle jerk reflexes were brisk (3+ bilaterally) with sustained clonus bilaterally. She had severe spasticity of her lower extremities and could not ambulate independently. She complained of urinary retention and had a post-void residual of 600 cc. Craniospinal imaging was performed.

Figure 2

Figure 2. CT revealed slit-like lateral and third ventricles (A) and a markedly dilated fourth ventricle (B), consistent with a functioning ventriculoperitoneal shunt and isolated 4 ventricle.

Figure 3

Figure 3. MRI flow-study of the brain demonstrated an occluded aqueduct (A) but open communication between the 4 ventricle and the cisterna magna. Post-gadolinium images revealed an enhancing lesion in the proximal fourth ventricle (B). C) Zoomed-in view of the suspected cysticercus occluding the distal aqueduct (white arrow).
Figure 4
Figure 4. MRI of the spine showed multiple cystic, intradural extramedullary lesions within the cervical, thoracic (A, B) and lumbar (C, D) spinal levels. Large cysts within the extramedullary spaces distorted the spinal cord and caused significant compression in the lower cervical and upper thoracic regions. Axial images (B, D) demonstratedystercerci within the cysts.

Treatment and Postoperative Course: She was taken to the operating room for suboccipital craniectomy, posterior fossa exploration and aqueductoplasty. We encountered and removed a small, cystic lesion from the distal aqueduct and proximal fourth ventricle and performed an aqueductoplasty using a ventricular catheter and Fogarty balloon dilatation.

Video 1. Narrated video showing the steps of the dissection, cyst removal and aqueductoplasty.

Postoperatively, the patient had complete resolution of her headaches, nausea and vomiting. Pathology, serological and CSF laboratory studies confirmed the diagnosis of NCC.

Figure 5
Figure 5. The specimen submitted for neuropathological examination revealed a cyst wall of a cysticercus. It consisted of a cuticular layer with hair-like protrusions, a middle cellular layer, and an inner reticular layer.

Postoperative MRI flow-study revealed a patent aqueduct and decrease in diameter of the fourth ventricle.

Figure 6
Figure 6. MRI showed complete removal of the enhancing lesion with the proximal 4 ventricle (A). The flow-study demonstrated a flow-void across the aqueduct confirming re-establishment of CSF flow between the 3 and 4 ventricles (B). The 4 ventricle also decreased in size (C).

She was also treated with high-dose corticosteroids for 8 weeks and a protracted course of albendazole. After 1 week of antiparasitic therapy and steroids, she evinced improvement in her gait and signs of myelopathy. She completed her course of antibiotics and continues to have improvement in her lower extremity coordination and strength. All spinal lesions have decreased in size and none have required laminectomy and removal by 9 months following initiation of antihelminthic therapy.

DISCUSSION
The dissemination of cranial NCC is in the cerebrum (91%), ventricles (6%), and subarachnoid space (2%). Approximately 15 to 54% of patients present with
ventricular and subarachnoid (cisternal) forms, which is more aggressive than parenchymal NCC. The racemose form of NCC accounts for 10% of cases and manifests as a single, large vesicle or as a mass of vesicles that may measure up to 10 cm.

Though the number of cranial NCC cases has been increasing, spinal NCC is relatively rare and occurs in only 1.6 to 13% of patients. The low incidence is attributed to the understanding that blood flow to the brain is 100-fold greater than blood flow to the spine. Alternatively, the lower incidence may be an underestimate due to the asymptomatic nature of small cysticercal cysts in the spinal canal.

Leptomeningeal (extramedullary) and intramedullary variants are the two forms of spinal NCC currently recognized. The former is 6 to 8 times more common and is thought to result from larvae migration from the cerebral to the spinal subarachnoid space. The latter is a result of hematogenous or ventriculoependymal migration. In the past, 30% of spinal NCC patients had intracerebral involvement while 25% of spinal NCC patients had intramuscular involvement. More recently, it has been estimated that isolated spinal cases are rare and that 75 to 100% of spinal NCC cases present with concurrent intracerebral involvement. The approximate distribution of spinal cysticerci is as follows: 35% cervical; 44.5% thoracic; 15.5% lumbar; 6% sacral. Spinal NCC is considered more aggressive than cranial NCC due to the relatively lower overall volume and cross-sectional area of the spinal canal compared to the intracranial space.

NCC may be treated medically with praziquantel or albendazole, with studies indicating albendazole as more clinically effective than praziquantel in achieving cyst reduction. However, cyst degeneration from medical therapy may result in a robust host inflammatory response, which may cause dangerous increases in intracranial pressure and spinal cord compression. Therefore, surgical treatment is often indicated in spinal NCC as a first-line treatment option and high-dose steroids are recommended during the acute phase of treatment.

Gravity and CSF flow patterns promote cyst migration to the fourth ventricle, causing a rare, late complication known as TFV. Similarly, Brun’s Syndrome may result from intermittent CSF obstruction from a ball-valve mechanism. Hydrocephalus in NCC cases is traditionally treated with a CSF shunt. More recently, however, neuroendoscopy has emerged as an alternative treatment in order to avoid shunt complications and failure.

Flexible cerebral endoscopy effectively treats Brun’s Syndrome and avoids the complications of a standard open craniotomy. Longatti et al. advocate endoscopy for its unique and superior visualization of neuroanatomy. A flexible neuroendoscope through a rigid scope into the fourth ventricle allows for successful excision of fourth ventricular cysts. Proaño and colleagues performed an observational, comparative cohort study for intraventricular and subarachnoid basal cisterns NCC and found that minimally invasive flexible endoscopic surgery provided effective and rapid removal of cysts and faster patient recovery. A retrospective study of 10 patients by Husain et al. determined that intra-fourth ventricular cyst transaqueductal removal with endoscopic third ventriculostomy using a rigid endoscope and catheter serves as an effective alternative to exploration of the posterior cranial fossa.

In the case described herein, the patient had an extremely steep tentorium and trajectory along the aqueduct. We believed such anatomy precluded a completely endoscopic, transventricular approach through the 4th ventricle. We chose an open approach to allow a wider view of the 4th ventricle and created a bend in the stylet that allowed an optimized trajectory through the aqueduct. Thorough evaluation of each patient’s intracranial anatomy is paramount for operative success, especially when considering lesions within the center of the brain and the feasibility of minimally invasive, endoscopic approaches.

CONCLUSION

We report a case of subacute thoracic myelopathy and acute deterioration from a TFV from aqueductal occlusion from untreated NCC. She underwent urgent posterior fossa exploration. A fourth ventricular lesion consistent with NCC was removed from the distal aqueduct and the aqueduct was opened via aqueductoplasty with a ventricular catheter and balloon dilatation. Intraoperative visualization and postoperative MRI flow-studies confirmed patency of the aqueduct and there was complete resolution of her fourth ventricle dilatation. With antiparasitic and corticosteroid therapy, her myelopathy has slowly improved over 9 months of follow-up without need for laminectomy thus far.

References

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