Intraventricular *Taenia solium* Cysts Presenting with Bruns Syndrome and Indications for Emergent Neurosurgery

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Abstract. Bruns syndrome is an unusual phenomenon, characterized by attacks of sudden and severe headache, vomiting, and vertigo, triggered by abrupt movement of the head. The presumptive cause of Bruns syndrome is a mobile deformable intraventricular mass leading to an episodic obstructive hydrocephalus. Intraventricular tumors have been associated with Bruns syndrome; however, few cases of intraventricular neurocysticercosis have been reported to present with Bruns syndrome. We report the first series of fourth ventricular neurocysticercosis presenting with Bruns syndrome in the United States and review the other published cases where surgery was indicated.

INTRODUCTION

Neurocysticercosis is caused by the larval form of *Taenia solium* (cysticerci) and is one of the few helminthic infections of the central nervous system. Humans become *T. solium* tapeworm carriers by ingesting undercooked pork containing cysticerci in muscle tissue. Once ingested, the scolex evaginates and attaches to the human small intestine, which later gives rise to the proglottids, and each proglottid segment contains 50,000 to 100,000 eggs. Most individuals with intestinal tapeworm infection do not develop symptomatic cysticercosis¹; however, are at risk for fecal–oral autoinoculation or fecal–oral transmission of eggs to other individuals and subsequent development of neurocysticercosis.

Neurocysticercosis is categorized on cysticerci location into parenchymal and extraparenchymal forms. Extraparenchymal forms include intraventricular, subarachnoid, intra-ocular, and spinal diseases. Cysticerci develop in the ventricular system as free floating cysts in the ventricular cavity or attached to the choroid plexus in 10% to 20% of patients presenting for clinical attention.^{2,3} Symptoms typically develop when cysticerci become lodged in the ventricular outflow tracts with consequent development of obstructive hydrocephalus and increased intracranial pressure.^{2,3}

The Bruns syndrome is an unusual phenomenon, characterized by attacks of sudden and severe headache, vomiting, and vertigo, triggered by abrupt movement of the head. The presumptive cause of Bruns syndrome is a mobile deformable intraventricular mass leading to an episodic obstructive hydrocephalus resulting from an intermittent or positional cerebral spinal fluid (CSF) obstruction with elevation of intracranial pressure due to a ball valve mechanism.⁴ Intraventricular neurocysticercosis (IVNCC) has been reported as an important etiology of Bruns syndrome.^{5–9}

Here we describe the cases of two patients who developed Bruns syndrome secondary to fourth ventricular neurocysticercosis and review 12 similar cases reported in the English literature^{5–9} (Table 1). Cases were identified in the literature by a PubMed search using terms “intraventricular neurocysticercosis” and “Bruns.”

CASE REPORT

Case 1. A 40-year-old woman, an immigrant from Nicaragua, presented to the hospital with a 1-month history of worsening headaches associated with severe nausea and vomiting; symptoms worsened with head movement. The patient was diagnosed with hydrocephalus 4 years prior, with similar symptoms; however, symptoms resolved at that time without any medications or interventions. A month before hospital admission she again had similar symptoms, a ventriculoperitoneal (VP) shunt was placed for the hydrocephalus with only partial resolution of symptoms. The patient immigrated to the United States at age 3 with no further travel or visitors from Nicaragua. Magnetic resonance imaging (MRI) of the brain showed a fourth ventricle cyst with extrusion into the right foramina of Luschka along with dilatation of the fourth ventricle (Figure 1A–C). These radiological findings were characteristic for IVNCC and the patient was started on treatment with albendazole 400 mg twice a day and dexamethasone 12 mg daily. Considering the obstruction and location of lesion, extraction of the cyst was strongly recommended and the patient underwent suboccipital craniotomy with removal of the cyst and scolex from posterior fossa. Pathology confirmed cystic calcified fibrovascular tissue in the specimen. Serum cysticercosis antibody was tested and was positive (6.08, reference range: < 0.90, Labcorp, Burlington, NC). Patient tolerated the surgery well with resolution of her symptoms and was discharged with a 4-week course of albendazole 400 mg twice a day and a tapering dose of dexamethasone (2 mg decrease each week). A follow-up computed tomography (CT) scan of the brain, 2 days after the surgery, demonstrated improvement in the size of the hydrocephalus (Figure 1D and E).

Case 2. A 39-year-old woman, an immigrant from Mexico, presented with an acute episode of syncope and an associated 7-month history of episodic headaches, dizziness, nausea, vomiting, and tinnitus associated with head movements. Six months before the current presentation, a CT scan was obtained, which showed stable mild hydrocephalus, a stable cystic fourth ventricular mass with a subtle soft tissue attenuation nodule, and evidence of prior granulomatous disease. MRI at the time of presentation showed a 1.3-cm irregular cyst in the fourth ventricle, resulting in moderate intratentorial and supratentorial hydrocephalus that had progressed since the prior CT scan and unchanged punctate nonspecific calcification in right superior frontal gyrus (Figure 2A–C). Because this cyst was partially obstructive, she underwent ventriculostomy with

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TABLE 1
Summary of cases with Bruns syndrome secondary to intraventricular neurocysticercosis

Case	Age (year), sex (reference)	Presentation	Duration	Diagnostic modality	Anatomical position of cyst	Country of origin	Treatment
1	40, F Case 1	Headache, nausea, vomiting	4 weeks	CT/MRI	Fourth ventricle	Nicaragua	Ventriculoperitoneal (VP) shunt, albendazole, dexamethasone, surgery
2	39, F Case 2	Headaches dizziness, nausea, vomiting, tinnitus, syncope	7 months	MRI	Fourth ventricle	Mexico	External ventricular drain, albendazole, dexamethasone, VP shunt
3	43, F ⁵	Headache, nausea, vomiting	4 weeks	MRI	Fourth ventricle	Brazil	Albendazole, dexamethasone, VP shunt, surgery
4	24, F ⁶	Headache, vertigo, vomiting	8 weeks	MRI	Fourth ventricle	India	Surgery
5	17, F ⁷	Headache, vomiting, syncope	2 weeks	CT	Third ventricle	India	Albendazole, endoscopic surgery, antiepileptic drugs
6	15, F ⁷	Headache, vomiting, syncope	4 weeks	CT	Third ventricle	India	Albendazole, endoscopic surgery
7	69, M ⁸	Headache, nausea, confusion	3 weeks	CT	Fourth ventricle	Thailand	Surgery-cyst removal
8	22, M ⁹	Headache, vomiting		MRI	Third ventricle	Mexico	Endoscopic surgery
9	31, F ⁹	Headache, papilledema		MRI	Third ventricle	Mexico	Endoscopic surgery
10	17, F ⁹	Headache, visual disturbance		MRI	Third ventricle	Mexico	Endoscopic surgery
11	28, M ⁹	Headache, vomiting		MRI	Third ventricle	Mexico	Endoscopic surgery
12	43, M ⁹	Lethargy		MRI	Third ventricle	Mexico	Endoscopic surgery
13	54, M ⁹	Headache, vomiting, diplopia		MRI	Third ventricle	Mexico	Endoscopic surgery
14	35, F ⁹	Headache, Parinaud sign		MRI	Third ventricle	Mexico	Endoscopic surgery

CT = computed tomography; MRI = magnetic resonance imaging.

external ventricular drain placement. Pathologic evaluation revealed incompletely necrotic metazoan tissue consistent with the diagnosis of neurocysticercosis. Serum cysticercosis antibody was also positive. Because of radiographic and clinical evidence of intraventricular disease, she was started on albendazole 400 mg by mouth twice daily and dexamethasone 6 mg daily. The patient continued to have intermittent episodes

of nausea, headache, and numbness of the right cheek. Steroids were increased to 12 mg per day in an attempt to resolve these symptoms. However, after multiple failed attempts to wean the external ventricular drain (Figure 2D and E), she underwent placement of a VP shunt on hospital day 17 and her hydrocephalus resolved (Figure 2F). She was discharged home on day 20 with plans to complete a 4-week course of albendazole

FIGURE 1. Magnetic resonance imaging (MRI) images at presentation. **A** and **B** show fourth ventricle cyst with invagination through right foramen of Luschka. **C** is a sagittal view showing extensive fourth ventricle involvement. Computed tomography (CT) 2 days after ventriculostomy. **D** shows decompression of cyst in fourth ventricle and **E** shows postsurgical changes and mild hemorrhage into right foramen of Luschka.

FIGURE 2. Patient 2: Magnetic resonance imaging (MRI) images at presentation. **A** and **B** show irregular obstructing cyst in fourth ventricle. Sagittal view T2 fast spin-echo (FSE) (**C**) with extensive lateral, third and fourth ventricle dilatation. (**D** and **E**) Computed tomography (CT) 9 days after ventriculostomy, increasing dilatation of lateral, third and fourth ventricles, external ventricular device tip in third ventricle. (**F**) CT 12 days after ventriculostomy with decreasing dilation and sulcal effacement, and ventriculoperitoneal (VP) shunt placement.

and on 60 mg prednisone daily with plans to taper after completion of a 30-day course.

DISCUSSION

In this case series, we describe the first two patients with IVNCC presenting as Bruns syndrome seen in the United States. Individuals with cysticercosis also present for medical attention outside of endemic areas, particularly where there are significant numbers of immigrants.¹⁰ In a prospective study of 1,800 patients presenting with seizures to 11 U.S. emergency departments over a 2-year period, neurocysticercosis was the etiologic agent in about 2% of cases. Further, neurocysticercosis was observed more frequently in emergency departments that had a higher proportion of immigrant Hispanic patients than the other hospitals.¹⁰ The reported patients in our series had remote history of travel to an endemic area, but none in over the last three decades. This highlights the fact that tapeworm carriers and most individuals with taeniasis remain at risk for fecal–oral inoculation of eggs and subsequent development of neurocysticercosis.

IVNCC could present with various features; however, life-threatening obstructive hydrocephalus caused by positional CSF obstruction (Bruns syndrome) is an infrequent and striking feature of the disease.^{4,9} Symptoms could be acute to subacute as highlighted in our series (duration of symptoms: 2–8 weeks) and could present as mild to severe symptoms with loss of consciousness.⁷ The manifestation of intraventricular cysts depends on the involved ventricle and is more severe if the fourth ventricle is occupied.¹¹ Further assessing for worsening symptoms of increased intracranial pressure with head movement could help clinicians make the association of Bruns syndrome.

The diagnosis of neurocysticercosis is multimodal in nature and includes pertinent clinical correlates and neuroimaging.

CT and MRI scan of the brain are used routinely in the diagnosis of IVNCC. Identification of a scolex in a cystic lesion is the pathognomonic radiographic finding. Scolices appear as rounded or elongated bright nodules 2–4 mm in diameter within the cyst cavity. Other radiographic findings suggestive of neurocysticercosis include hydrocephalus and leptomeningeal enhancement.¹² MRI is the preferred modality for diagnosis, as the MRI signal properties of the cystic fluid or the scolex differ and inflammation is marked with hyper-intensity signals, allowing for reliable diagnosis.¹² Pathology, if available, helps confirm the diagnosis. Typically, membranous structures filled with fluid or at times, scolices (composed of rudimentary bodies and heads with suckers and hooks), are seen on histopathology.³ Serological analysis has an adjunct role in supporting the diagnosis; however, delaying treatment while awaiting results is not recommended in cases that require urgent diagnosis.⁷ The test of choice for antibody detection is the enzyme-linked immunoelectrotransfer blot assay (EITB) developed by the Centers for Disease Control.^{13,14}

Treatment of IVNCC should be personalized, depending on the location and the severity of the disease. Medical management should be avoided for patients presenting with neurological emergency or increased intracranial pressure. Surgical relief of obstructive hydrocephalus should be the priority. Use of a VP shunt or a temporary ventriculostomy for CSF diversion to relieve the obstruction has been used in emergent situations.¹¹ The most common cause for dysfunction of the VP shunt is obstruction of the shunt by the gelatinous material from cysts or the high CSF protein level, indicating removal of the cyst as a definitive treatment modality. Cysticerci can be resected either with an endoscopic or open approach. Endoscopic approach may be used for cysts in the lateral and third ventricles⁹; however, extraction of the cysticerci from the fourth ventricle may be surgically challenging and could require open resection.

Anthelmintic drugs namely albendazole and praziquantel have reasonable efficacy in all forms of neurocysticercosis. However, the role of anthelmintic therapy in IVNCC remains debated. The use of anthelmintics and steroids are recommended to eradicate residual viable cysts and to decrease the incidence of shunt failures after surgical removal of the intraventricular cyst.¹⁵ Albendazole is preferred over praziquantel in IVNCC because of its superior efficacy and absence of drug interactions with corticosteroids and antiepileptic drugs. A recent trial suggests that high-dose albendazole (30 mg/kg/day) increases clearance of subarachnoid and intraventricular cysts as compared with conventional dosing (15 mg/kg/day).¹⁶

When the *T. solium* dies or cyst ruptures, liberation of antigenic substances triggers host immune response resulting in an inflammatory response throughout the ventricular system.¹⁷ No controlled data are available on the use of corticosteroids in IVNCC; however, steroids should be considered to reduce the incidence of inflammation and host-mediated response since they have been shown to be efficacious in reducing inflammation in other forms of neurocysticercosis.¹⁸

It should be noted that surgery with cyst removal was the definitive treatment of the reported patients. In the literature, three of the 14 (21%) patients had VP shunts placed; however, two of them required subsequent surgeries. For patients with conservative management including cysticidal medications and/or placement of VP shunts, there was high risk of symptom recurrence as demonstrated in the reported patients. Careful monitoring of these patients is required to prevent a recurrence of severe obstruction.

In this case series we describe the first reported patients of IVNCC presenting as Bruns syndrome seen in the United States. Individuals with neurocysticercosis also present for medical attention outside of endemic areas, particularly where there are significant numbers of immigrants.¹⁰ These cases highlight that neurocysticercosis should be considered if risk factors and neurological symptoms varying from mild to severe, including loss of consciousness, exist in a patient presenting with an obstructive hydrocephalus. VP shunt or a temporary ventriculostomy for CSF diversion should be used to relieve the obstruction in emergent situations. Surgical or endoscopic removal of the cyst appears to be the definitive management of such cases.

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