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Fourth ventricle neurocysticercosis presenting with Bruns' syndrome: A case report

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Case Report

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ABSTRACT

Background: Neurocysticercosis (NCC) is the most common helminthic infection of the central nervous system. We present a case of the fourth ventricle NCC that presented with Bruns' syndrome (with headaches worsened by head movements, gait ataxia, and vomiting) and the operative technique used for cyst removal.

Case Description: A 39-year-old woman was admitted to the emergency department with moderate-to-severe headaches that got worse in the afternoon and were triggered by head movements, relieved on prone position, and were associated with gait ataxia and vomiting. A brain magnetic resonance imaging (MRI) showed a fourth ventricle cyst, with an eccentric enhancing nodule compatible with a larvae scolex and associated obstructive hydrocephalus. The patient was positioned prone and underwent a microsurgical resection of the cyst without rupture through a telovelar approach to the fourth ventricle. The postoperative MRI confirmed complete cyst removal and resolution of the hydrocephalus. At 12-month follow-up, the patient remains with no signs of disease recurrence.

Conclusion: In this report, we depict a case of intraventricular NCC successfully treated with a single surgery, allowing intact cyst removal and achieving effective hydrocephalus treatment with no need to resort to cerebrospinal fluid diversion techniques.

Keywords: Bruns' syndrome, Hydrocephalus, Intraventricular neurocysticercosis, Telovelar approach

INTRODUCTION

Neurocysticercosis (NCC) is the most common helminthic infection of the central nervous system.^[14] It is caused by the larval stage of *Taenia solium*. The disease has two forms: intraparenchymal and extraparenchymal.^[7] Intraparenchymal NCC is an important cause of seizures.^[4,5] Extraparenchymal NCC is a major cause of hydrocephalus.^[4]

The disease is endemic to low-income countries where proper sanitation is unavailable. However, high-income regions are not NCC-free mainly due to immigration from endemic areas.^[4] In this report, we focus on extraparenchymal intraventricular NCC, presenting the case of a patient with a fourth ventricle cyst, treated with a single surgical procedure that resolved both the infection and hydrocephalus. The patient presented with severe headaches triggered by head movements associated with gait ataxia and vomiting – the triad of Bruns' syndrome.^[8]

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CASE REPORT

A 39-year-old woman from Cape Verde was referred to the emergency department with severe headaches triggered by head movements, gait ataxia, and vomiting. The clinical picture had a protracted course and had been worsening in the past few months. Neurological and laboratorial evaluation was unremarkable.

The brain magnetic resonance imaging (MRI) showed a fourth ventricle cyst, 2.6×1.8 cm (cephalocaudal \times transverse axis) with an enhancing nodule compatible with a scolex and associated obstructive hydrocephalus [Figures 1 and 2]. She repeated the MRI for surgical planning with a T2-weighted sagittal sequence. In the second MRI, the cyst and its content showed a slightly distinct morphology and a different position in the fourth ventricle, suggesting a mobile behavior.

We devised a surgical approach to achieve both cyst removal and hydrocephalus treatment. The patient was placed in the prone position with the head supported by a three-point head holder. The fourth ventricle was accessed through a median suboccipital craniotomy and telovelar approach [Figure 3]. The cyst was identified and dissected from the ependyma, allowing for *en bloc* removal without rupture. The cerebrospinal fluid (CSF) flow was restored. We performed a watertight dural closure and a bone flap cranioplasty with a double-sided clamp system.

The postoperative period was uneventful and the patient was discharged to the ward from intensive care in the first 24 h. Postoperatively, she fully recovered from the presenting symptoms over 72 h. MRI showed complete cyst removal and no complications [Figure 4]. She was discharged home on day 7 after surgery. Histological analysis confirmed NCC.

The patient repeated an MRI at 1-, 6-, and 12-month follow-up with no signs of recurrence.

DISCUSSION

Extraparenchymal NCC is associated with high mortality due to hydrocephalus.^[4] Incidence peaks between the third and fourth decades. Del Brutto's diagnostic criteria help identify NCC. Absolute criteria include histological characterization and cystic lesions with scolex on neuroimaging.^[3]

In 12% of patients, the disease presents with hydrocephalus, caused by blockage of CSF flow through the ventricles, occlusion of foramina of Magendie and Luschka, or inflammatory reaction causing aqueduct stenosis.^[9] We describe a patient that presented with the unusual Bruns' syndrome, caused by a mobile intraventricular lesion leading to episodic bouts of acute hydrocephalus.^[17] This syndrome also occurs with the third ventricle cysts and tumors but was originally described in the fourth ventricle NCC.^[1]

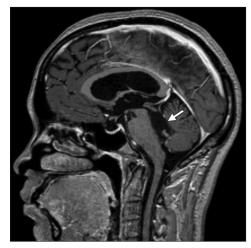


Figure 1: Preoperative sagittal T1-weighted gadolinium-enhanced magnetic resonance imaging showing a cyst (arrow) in the fourth ventricle with a tubular hyperintense structure.

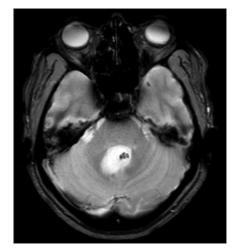


Figure 2: Preoperative axial T2-weighted fast field echo showing the hypointense four cup-shaped suckers of the scolex.

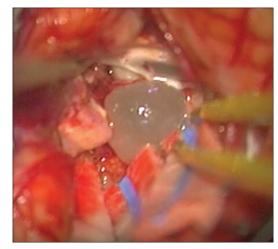


Figure 3: Intraoperative morphology of the lesion.

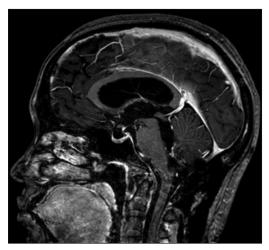


Figure 4: Postoperative sagittal T1-weighted gadolinium-enhanced magnetic resonance imaging showing cyst removal.

Differential diagnoses of intraventricular cystic lesions comprise infectious, neoplastic, and nonneoplastic cysts.^[19] Infectious cysts other than neurocysticercosis include tuberculomas, pyogenic abscesses, hydatidosis, and toxoplasmosis.^[10,21] Neoplasms that can present as cystic lesions in this location include ependymoma, subependymoma, and choroid plexus tumors.^[11] Arachnoid cysts, Blake's pouch cysts, ependymal cysts, and colloid cysts are well-known nonneoplastic/noninfectious cysts that can present in the fourth ventricle.^[19] Medical history and epidemiology can help to define the likelihood of a diagnosis. In this case, the presence of the scolex inside the cyst and the epidemiological context of the patient made NCC the first diagnostic hypothesis.

Therapeutic approaches include surgery and medical treatment (corticosteroids and antihelminthics).^[13] We chose an open approach to remove the cyst, with a telovelar approach to the fourth ventricle with successful lesion removal. The endoscopic approach to the fourth ventricle, although described with favorable results, involves the passage of a flexible endoscope through the aqueduct, with the risk of major neurological deficit.^[6,18] There is a reported series in which 40% of patients treated with suboccipital craniotomy developed delayed hydrocephalus, needing a ventriculoperitoneal shunt.^[15] We highlight that our patient was successfully treated with a single surgery, avoiding shunt placement. This is relevant due to the high shunt failure rates in NCC.^[16] In a published report, 43.5% of patients with intraventricular NCC needed a shunt, with a failure rate of 40%.^[12]

The medical treatment for intraventricular NCC remains controversial. Antihelminthics are recommended in parenchymal NCC but have no clear benefits in intraventricular NCC when surgery is feasible with intact cyst removal.^[2,20] We followed the 2017 guidelines by the Infectious Diseases Society of America and the American Society of Tropical Medicine and Hygiene which state that in case of isolated cyst removal without rupture, antihelminthics are not recommended but corticosteroids are useful to reduce brain edema in the perioperative period.^[20]

CONCLUSION

Intraventricular NCC remains a surgically treated disease. Herein, we depicted a case of a patient presenting with an unusual type of obstructive hydrocephalus – Bruns' syndrome. In this case, the cyst location at the fourth ventricle strategically blocked CSF flow and its removal resolved the hydrocephalus. The patient was successfully treated with a single surgical approach and a ventriculoperitoneal shunt placement was avoided.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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