

CASE REPORT

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Intraventricular neurocysticercosis presenting as Bruns' syndrome: An uncommon presentation

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Abstract

Background: Though parenchymal neurocysticercosis is common and a major contributor to burden of seizures in most parts of the world, intraventricular neurocysticercosis (IVNCC) comprises 10–20% of cases and poses a diagnostic challenge to the clinician.

Case presentation: We report an adult female presenting with intermittent occipital headache, used to be worse in lying down position, and aggravated with head movements, and there was mild relief in the sitting position. Her physical examination was unremarkable, and laboratory tests were within normal limits. Her multimodal neuroimaging showed cystic lesion in the fourth ventricle suggestive of neurocysticercosis. Patient underwent neuroendoscopic removal of the cyst, and the final diagnosis was confirmed on histopathology. Post removal of cyst patient had complete resolution of her symptoms.

Discussion: Intraventricular neurocysticercosis can present as acute hydrocephalus which may clinically manifest as Bruns' syndrome in which sudden attacks of headache vertigo and nausea or vomiting are precipitated by abrupt head movements which was observed in our patient. Multimodal neuroimaging supported by histopathology helped in confirmation of the diagnosis, thus averting an inadvertent use of unnecessary medications in such patients. Furthermore, neuroendoscopy has evolved as minimally invasive technique for extirpation of fourth ventricular cysts.

Keywords: Intraventricular cysticercosis, Multimodal imaging, Hydrocephalus, Cystic lesion, Neuroendoscopy

Background

Neurocysticercosis poses a complex diagnostic and management dilemma in view of its varied presentation and is often confused with tuberculomas or malignant cystic lesions. Factors determining immediate management include whether patient is symptomatic, location of cysts, and the host response. Parenchymal location for these cysts is common; however, cysts can also be seen in the ventricular system though uncommon. The natural evolution of intraventricular cysts is limited by their clinical expression as the cysts which block the cerebrospinal

flow cause obstructive hydrocephalus and the ones which are attached to the ventricular wall involute and eventually resolve. Whether the intraventricular cysts require medical or surgical intervention would depend on certain typical clinical clues, thus posing a diagnostic challenge to the clinician. We present a patient with Bruns' syndrome, caused by a cystic lesion in the ventricular system confirmed by multimodal imaging and attained complete recovery after treatment, and nature of lesion was proven by histopathology, thus requiring a multidisciplinary team for effective management.

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Case presentation

A 32-year-old lady presented to our neurology emergency with 1 month history of intermittent episodes of occipital headache, which was insidious onset, used to be worse in lying down position with mild relief in sitting position and aggravated with abrupt head movements. She did not report any seizures, nausea, photophobia, vision loss, dizziness, fever, loss of consciousness, or trauma. Her physical and neurological examination was unremarkable. Her computed tomography (CT) scan (16 slice GE medical systems Optima CT 540) revealed dilated lateral, 3rd and 4th ventricle but underlying cause for obstructive hydrocephalus could not be demonstrated. Multimodal magnetic resonance imaging (MRI) brain (Philips, Ingenia 3 Tesla MR system, India) and axial sequences (Fig. 1a, b) showed cystic lesion in 4th ventricle. Constructive interference in steady state 3 dimensional (CISS3D), a new T2 weighted high resolution sequence (Fig. 1c–e—red arrow), was used to demonstrate the cyst in 4th ventricle, and susceptibility weighted image (SWI) showed a small focus of blooming (Fig. 1f—blue arrow) suggestive of calcification. A provisional diagnosis of intraventricular neurocysticercosis (IVNCC) was considered. Patient underwent neuroendoscopic removal of the cyst, and

histopathological examination showed neurocysticercal cyst with characteristic three layered wall, which confirmed the diagnosis (Fig. 2). Post neuroendoscopy patient had complete resolution of her symptoms and neuroimaging was normal (Fig. 3).

Discussion

Neurocysticercosis is a common parasitic neurologic disease worldwide, but in some parts of the world, it remains relatively rare, and the presentation is variable depending on the location of cyst. It is caused by the larvae of pork tapeworm (*Taenia solium*) and acquired from a human carrier through faecal-oral route. Approximately, 10–20% of patients worldwide [1] are present with IVNCC, which has a high mortality rate owing to secondary obstructive hydrocephalus. Viable cysts are freely mobile and may lodge anywhere in the foramina or the aqueduct resulting in mechanical obstruction or as a result of arachnoiditis in chronic cases [2]. The cysts reach the ventricular space through the choroid plexus and are more frequently found in the fourth ventricle, likely due to gravitational forces which favor migration from supratentorial ventricles or may directly enter through the choroid plexus. The size of the cyst and the ventricular foramina could also be one

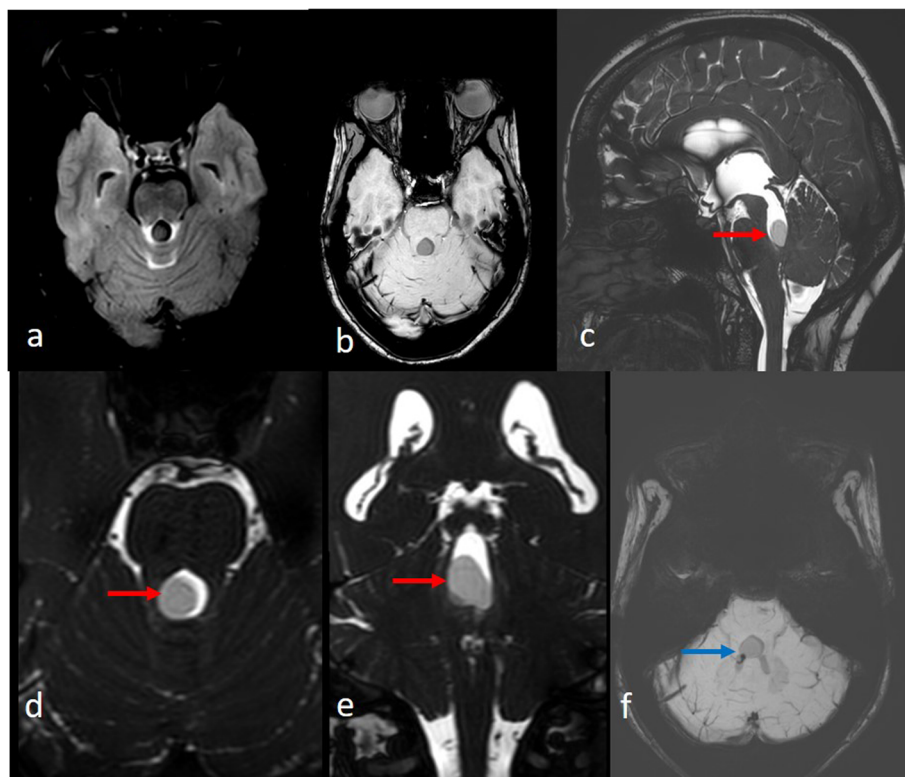


Fig. 1 Multimodal magnetic resonance imaging brain axial images (a, b) showing cyst in the 4th ventricle, constructive interference in steady state 3 dimensional CISS3D sequence-sagittal, axial, and coronal (c–e; red arrow) delineating the cyst in 4th ventricle, and there is evidence of blooming on susceptibility images (f; blue arrow)

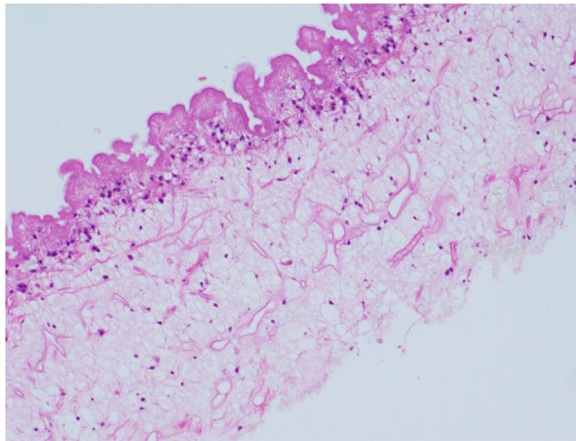


Fig. 2 Neurocysticercal cyst showing the characteristic three layered wall: a cuticular layer with hair-like microtrichia, middle cellular layer, and an inner reticular layer with focal calcification

of the factors responsible for frequent involvement of fourth ventricle. The presenting features of hydrocephalus are headache, nausea, vomiting, altered sensorium, and papilledema. Acute hydrocephalus may present as Bruns' syndrome in which sudden attacks of headache, vertigo and nausea, or vomiting were precipitated by abrupt head movements which was present in our

patient [3]. Episodic hydrocephalus in Bruns' syndrome is due to intermittent obstruction of cerebrospinal fluid flow by ball valve movement of intraventricular cysts. Mechanism of hydrocephalus is either ventricular obstruction or arachnoiditis (either basal or spinal). Bruns' syndrome has been originally described in fourth ventricular NCC but has also been reported in other lesions like third ventricular lesions [4, 5]. Lu Zhengqi and colleagues described the first case of Bruns' syndrome secondary to tuberculoma which mimicked neurocysticercosis and the patient improved on treatment. Such cases especially a single cyst as seen in our case pose a diagnostic dilemma for a clinician and if misdiagnosed as tuberculoma can result in unnecessary side effects of antituberculous drugs. Though serological tests are available for diagnosis, they have a poor sensitivity and specificity. So, majority of cases are diagnosed with help of imaging; however, histopathology is the gold standard for diagnosing NCC. These intraventricular cysts are poorly defined on CT Brain and so routine MRI Brain is used for diagnosis. However, routine MRI sometimes may also not be able to delineate the nature of lesion; as result, newer imaging modalities like constructive interference in steady state 3 dimensional (CISS3D) and fast imaging employing steady-state acquisition (FIESTA) sequence promise improved detection of IVNCC in terms of better sensitivity [6]. In addition, previously

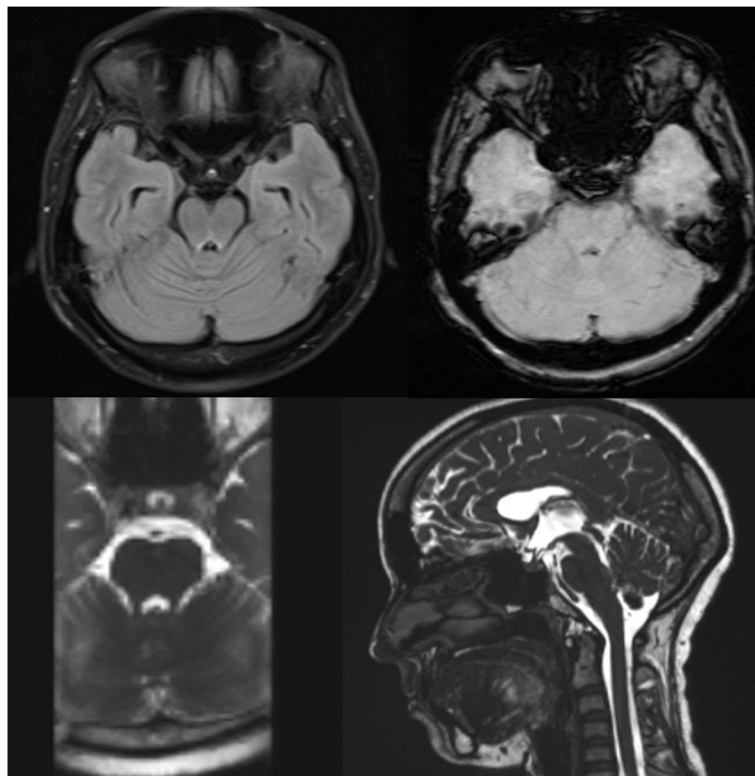


Fig. 3 Post neuroendoscopy neuroimaging showing normal fourth ventricle with no evidence of cyst

missed cases have been diagnosed using 3D CISS sequence [7]. These advanced imaging techniques have also been strongly recommended by Infectious Disease Society of America (IDSA) and the American Society of Tropical Medicine and Hygiene (ASTMH) [8]. Many conditions can cause hydrocephalus and thus mimic extra-parenchymal NCC. These include colloid cysts, cerebellar cystic hemangioblastoma, and tuberculomas. Treatment of IVNCC includes managing hydrocephalus and removal of the cyst. Flexible neuroendoscopy has emerged as the surgical approach for extirpation of the cysts, though surgical removal of fourth ventricular cysts with or without shunt surgery is recommended in cases where endoscopic removal is not possible [8, 9]. Accurate diagnosis is essential in such cases as it can prevent morbidity and mortality due to unnecessary interventions like lumbar puncture, shunt procedures, or initiation of antitubercular drugs without confirming the final diagnosis.

Thus, to conclude diagnosis of IVNCC can still pose a challenge despite modern neuroimaging methods because of poor specificity of clinical and imaging findings, a histopathological confirmation may be required to rule out a malignant lesion.

Abbreviations

IVNCC: Intraventricular neurocysticercosis; CISS3D: Constructive interference in steady state 3 dimensional; CT: Computed tomography; MRI: Magnetic resonance imaging; SWI: Susceptibility weighted image; FIESTA: Fast imaging employing steady-state acquisition; IDSA: Infectious Disease Society of America; ASTMH: American Society of Tropical Medicine and Hygiene

Acknowledgements

Not applicable.

Authors' contributions

All authors contributed equally for the manuscript. The authors read and approved the final manuscript.

Funding

Not applicable.

Availability of data and materials

The datasets supporting the conclusions are included in the article and are available with the corresponding author on reasonable request.

Machines used in the study computed tomography (CT) scan (16 slice GE medical systems Optima CT 540) and Magnetic Resonance Imaging (MRI) brain (Philips, Ingenia 3 Tesla MR system, India).

Ethics approval and consent to participate

We confirm that the ethical clearance was not required for publication of this case report. Written consent was obtained from the patient.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Competing interests

The authors declare that they have no competing interests.

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Received: 19 December 2019 Accepted: 28 May 2020

Published online: 17 June 2020

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