Case Report

An Unusual Case of Neurocysticercosis

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Abstract
Neurocysticercosis (NCC) is a significantly neglected tropical disease and, with increasing globalisation, a notable emerging infection in the developed world. It is one of the seven neglected endemic zoonoses targeted by the World Health Organization and is considered a common infection of the nervous system caused by the Taenia solium. It is known to be the primary cause of preventable epilepsy in many developing countries. Neurocysticercosis is a specific form of the infectious parasitic disease cysticercosis which is caused by infection with Taenia Solium, a tapeworm found in pigs. Neurocysticercosis commonly occurs by the ingestion of Taenia solium eggs after consuming undercooked pork, or contaminated water. The cysts formed by the infection grow within the brain and spinal cord within the nervous system causing neurologic symptoms such as dizziness, severe headache, epileptic seizures beside other pathological manifestations. It has been called a "hidden epidemic and "arguably the most common parasitic disease of the human nervous system". Clinical investigations are focused on development of effective treatments and reduction of side-effects induced by treatment, such as seizures, hydrocephalus, infarcts, and neuroinjury. A patient came to our ER with complaints acute onset of loss of consciousness for 1 day. The computed tomography and magnetic resonance imaging scans indicated multilobulated cystic mass with typical STARRY SKY APPEARANCE in the brain with the suspicion of neurocysticercosis

Keywords: Neurocysticercosis (NCC), zoonosis, STARRY SKY APPEARANCE.

Introduction
Neurocysticercosis (NCC) is the most common single cause of seizures/epilepsy in India and several other endemic countries throughout the world. Most importantly, it is one of the few diseases that can be eradicated, an accomplishment that would prevent millions of cases of epilepsy. Therefore it is of utmost importance to bring forth increase awareness about this disease in general population for prevention, early detection and seeking early treatment. It is also necessary for the clinicians to know about the various presentation of the disease across the globe for fast and effective diagnosis and treatment. Recent Indian studies using neuroimaging techniques suggest that the disease burden in India surpasses many other developing countries. Hence it is important to know the epidemiology, pathogenesis and diagnostic criteria
so as to assess the disease burden and adopt interventional strategies for its control.

**Case Presentation**

The patient 55 years old male, was brought to ER with C/O of loss of consciousness for last 24 hours, following binge alcohol intake of 1 day, he was found in an unconscious state at his home. Then he was taken to some hospital where dextrose containing fluid was infused but he did not regain consciousness so he was brought to our ER. No H/O Head injury or long term fever in recent past. No recent or past history of seizure/dogbite/vaccination.

**On Admission:** B.P: 140/80 mm of Hg, PR: 76/min, blood sugar: 320, ABG analysis: normal.

**On Examination:** Pt is partially awake & is responding to painful stimuli with only eyelid movements.

**CNS Examination: Higher Mental Function:** Pt is in a persistent vegetative state (i.e Respiratory function, sleep cycle is intact but there is no movement of any limbs, no awareness, complete loss of speech.

**Cranial Nerves:** 1st, 5th, 8th, 11th and 12th, could not be tested

2nd-pupils-B/L symmetrical, normally reacting to light.

3rd, 4th and 6th-ocular movements- only random eye ball movements, no movements in response to stimuli.

7th-no significant asymmetry over face

9th, 10th-gag reflex- absent.

**Spinomotor System: Bulk:** no obvious wasting of muscles in any of 4 limbs, Tone: ↓↓ in all 4 limbs

**Power:** no movements of any limbs to painful stimuli.

**Reflexes:** plantar-B/L extensor

**Abdominal reflex:** absent

**DTRs:** ↓↓ in all 4 limbs.

No involuntary movements.

Gait & Coordination could not be tested.

There is loss of bladder & bowel control.

Skull and spine examination is normal.

**Sensory System:** No movement of any limbs to painful stimuli.

**Investigations**

All the routine investigations including Complete Blood Counts, Renal function tests, Liver function tests were normal.

CSF analysis was also normal.

NCCT brain- Multiple NCC noted in B/L brain parenchyma, also involving the brain stem.

![NCCT brain](image1.png)

**MRI Brain:** Multiple lesion noted in B/L brain parenchyma with eccentric scolex in T1 sequence. We find so many cases of CNS NCC where one or few cysts can be seen in MRI brain but in this case there were extremely high number of cysts with typical STARRY SKY APPEARANCE.

![MRI brain](image2.png)

**Differential Diagnosis**

**Infective**

Multiple Tuberculoma

Multiple abscess

Histoplasmosis

Actinomycosis

Cryptococcosis
Toxoplasmosis
Neoplasm
Multiple metastasis
CNS lymphoma

**Inflammatory-Demyelinating Disorders**
Multiple sclerosis
Acute disseminated encephalo-myelitis
Neurosarcoidosis
Systemic lupus erythematosis

**Treatment**
He was treated with IV methylprednisolone and mannitol to decrease oedema, IV Eptoin to prevent seizures and other supportive therapies.

**Outcome and Follow up**
The patient remained in the same state for 40 days and then he expired.

**Discussion**
Neurocysticercosis is the result of accidental ingestion of eggs of *Taenia solium* (ie, pork tapeworm), usually due to contamination of food by people with taeniasis. *T solium* has a 2-host biologic cycle, with humans as the definitive hosts carrying the intestinal tapeworm, and pigs as the normal intermediate hosts harboring the larvae or cysticerci. This parasite has a head (scolex) with 4 suckers and a double crown of hooks, an unsegmented neck, and a large body with several hundreds of hermaphrodite proglottids. Cysticerci are ingested by humans through poorly cooked infected pork. Cysts evaginate in the small intestine, attach to the wall by its suckers and hooks, and develop strobila or chains of proglottids. From the distal end of the strobila, fertile eggs are excreted into the gravid proglottids. Up to 60,000 eggs may be contained in a proglottid. Pigs ingest stool contaminated with *Taenia* eggs, the embryos actively cross the intestinal wall, get into the bloodstream, and are transported to most tissues, where they reside as cysticerci. Larvae are found most commonly in the central nervous system (CNS), but they can also be located in the eye, muscle, or subcutaneous or other tissues.

Neurocysticercosis, caused by the larval form of the cestode *Taenia solium*, is associated with lack of sanitation, poor hygiene and free roaming pigs. Local transmission of the disease is, however, only possible in the presence of an adult *Taenia* carrier in the gut. Taeniasis is caused only by the consumption of pork infected with cysticercosis, but NCC can also occur in vegetarians and non-pork eaters.

The prevalence of neurocysticercosis as a cause of active epilepsy in India was calculated to be one per 1000 population. Thus, at least 1.2 million persons in India are suffering from active epilepsy due to neurocysticercosis[3]. It is the single most common cause of community acquired active epilepsy; 26.3% to 53.8% active epilepsy cases in the developing world including India and Latin America. The most common form of the disease in India was the solitary cysticercus granuloma (SCG) (first identified in 1989) which was seen in up to 60 per cent of patients with NCC[3,4]

Overall, seizures are the most common clinical presentation of symptomatic neurocysticercosis, with a reported prevalence of 70–90%. Headache is estimated to occur in over one-third of patients.[5] Focal neurological signs typically follow a subacute course similar to other space-occupying lesions. Less commonly, acute focal deficits can occur when inflammatory changes in penetrating arteries result in ischaemic stroke[6]. Signs of
increased ICP occur in over 10% of patients. All other manifestations—including chronic meningitis, encephalitis, changes in vision, pain in a nerve root distribution, or sensory changes—occur in <10% of patients. The clinical course of NCC is determined by both the anatomic location of the lesion and host inflammatory response. Isolated lesions in the brain parenchyma are common and associated with seizures when symptomatic.

The recognition of neurocysticercosis in acute setting is complicated by its pleomorphic clinical presentation and in some cases latent period after exposure can persist for years. This patient came with acute onset of loss of consciousness with the background of binge alcohol intake with no other relevant history creating problem for early diagnosis and treatment. It is therefore important to have a broad-mindedness during treating all cases.

The diagnosis of neurocysticercosis requires a combination of clinical evaluation, imaging and laboratory methods. Definitive diagnosis is rarely obtained and only accomplished by autopsy, biopsy or visualisation of the distinctive scolex on imaging. Neuroimaging can determine the location and stage of the lesions. The appearance of parenchymal cysts progresses from non-enhancing (if the lesion is viable with little host inflammatory response) to ring enhancing (when the cyst degenerates and triggers an immune response) to a calcified nodule or complete resolution. CT is more sensitive for the most common lesions (calcifications) and is more widely available in endemic countries, but MRI is more sensitive for all non-calcified forms of the disease.

Of the serological investigations, tests that detect anticysticercal antibodies are generally favoured over those that detect cysticercal antigens. The enzyme-linked immunoelectrotransfer blot (EITB) is preferred due to the high specificity (100%) and sensitivity (94–100%), if performed on serum. The EITB has much lower sensitivity for calcified cysts, particularly if a single lesion is present. An enzyme immunoassay is also available, but features a lower diagnostic sensitivity and yields more false positives due to cross-reactivity with Echinococcus. While fairly sensitive, antibody tests have limited specificity to detect active infection because antibodies may remain in the serum after parasites are no longer alive in the brain or as a result of soft tissue infection. In this case, the use of the ELISA, the test available in our laboratory at the time, may account for the false negative serum result. CSF serological testing (antibody or antigen) may be a useful adjunctive test, as was performed in this case, particularly if serum testing is not diagnostic. The diagnosis in this case required both, neuroimaging and laboratory investigations, highlighting the importance of multiple modes of evaluation combined with clinical reasoning in these cases.

The goals of treatment for neurocysticercosis include symptom control, eradication of the parasite and mitigation of the host's inflammatory process. Seizures are typically responsive to first-line antiepileptic drugs (phenytoin, carbamazepine or valproic acid). The ideal duration of seizure prophylaxis is unknown, but expert consensus recommends at least 2 years of treatment after the last seizure, followed by gradual tapering (except for patients with a single lesion that resolves without calcification). Other therapies to consider for symptom alleviation include anti-inflammatories, analgaesics and agents or interventions to manage intracranial hypertension.

The optimal treatment varies by location of the parasite. For viable parenchymal disease, antiparasitic medications increase the speed of radiographic resolution and decrease seizure recurrence. Albendazole (at 15 mg/kg/day by mouth for 8–15 days) is slightly more efficacious than praziquantel (at 50–75 mg/kg/day by mouth for 15 days). There is minimal safety information available for high-dose (30 mg/kg/day) albendazole treatment strategies, which was chosen in this case due to expert recommendation at the time of the patient's presentation. The combined use of albendazole and praziquantel at the recommended doses have been shown to have greater efficacy than either agent alone. The ideal treatment regimen for a solitary lesion remains uncertain, but
Antiparasitic therapies appear to modestly improve radiographic appearance and decrease seizure recurrence\(^{[20]}\).

Administration of antiparasitics can transiently worsen neurological symptoms due to antigen release and inflammation. Experts recommend the use of adjunctive corticosteroid therapy, a practice supported by preliminary studies showing it decreases seizure recurrence and increases resolution of lesions\(^{[21]}\). Dexamethasone (0.1 mg/kg/day) is the first-line treatment, initiated 1 day before antiparasitic therapy and continuing for 1–2 weeks followed by a slow taper\(^{[22]}\). In this case antiparasitics was not administered because of the suspicion of encephalitis in patient.

In summary, neurocysticercosis is an important cause of neurological disease in endemic regions and, increasingly, worldwide. The presentation of the disease is pleomorphic, requiring a combination of clinical evaluation, imaging and laboratory investigations for diagnosis. Because the latent period of the parasite can persist for years and risk factors for degeneration of viable lesions are not well characterized, a thorough history is fundamental in calibrating clinical suspicion. Given the rising prevalence outside of endemic regions, NCC is an important diagnostic consideration for a range of neurological presentations in an increasingly accessible world.

**Learning Points**

- Neurocysticercosis (NCC) is a preventable cause of severe neurological illness. Most importantly, it is one of the few diseases that can be eradicated, an accomplishment that would prevent millions of cases of epilepsy.
- The presentation of neurocysticercosis is pleomorphic, with varied signs and symptoms, some times manifesting acutely and sometimes may have a latent period of several years. Thus broad mindedness and knowledge about it is required during assessment of each case.
- Early diagnosis and treatment is the mainstay for decreasing morbidity and mortality of this disease.
- Diagnosis of neurocysticercosis requires a combination of clinical evaluation, imaging and laboratory methods. MRI is more sensitive for all lesions except the calcified forms, which are better evaluated with CT. Enzyme-linked immunoelectrotransfer blot is the serological test of choice. Because there is no single definitive test, a high level of suspicion should be maintained for neurocysticercosis.

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