

Reversible dementia as a presenting manifestation of racemose neurocysticercosis

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Abstract

Racemose cysticercosis is a less frequent presentation of neurocysticercosis (NCC). It's presentation and management is quite different from cerebral parenchymal NCC. Diagnosis of racemose cysticercosis is based on the combination of clinical, epidemiologic, radiographic, and immunologic information. Compared with cysticercus cellulose, which most commonly presents as seizures, racemose NCC due to its extraaxial location presents with raised intracranial pressure and meningitis, and frequently requires neurosurgical intervention. Dementia as a sole presenting feature of NCC is rare. We report a case of racemose NCC with dementia as the presenting manifestation. The outcome of dementia patients with NCC seems favorable in most cases therefore a high index of suspicion for NCC should be kept especially in endemic areas.

Key Words

Cisternal neurocysticercosis, intraventricular neurocysticercosis, neurocysticercosis, racemose neurocysticercosis, subarachnoid neurocysticercosis

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Introduction

Neurocysticercosis (NCC) is one of the most common helminthic infections of the central nervous system.^[1] *Cysticercus* refers to the resting stage of the larva of the cestodes belonging to the order Cyclophyllidea. In the intermediate host, it develops into a "bladder worm" consisting of a hollow vesicle with invaginated scolex on its wall and a central cavity containing fluid. The cerebral parenchyma is the most frequently affected central nervous system site. However, NCC can also occur in other extraparenchymal locations like intraventricular, subarachnoid/cisternal or mixed.^[2] The subarachnoid/cisternal form of NCC and majority of the intraventricular NCC are of racemose type, and differ from more common *cysticercus cellulose* in that they are larger, appear as multiloculated cysts and lack scolex.^[3] This intraventricular and subarachnoid (cisternal) form of NCC is seen in 15-54% of patients, and presents clinically in a more aggressive manner as compared with the parenchymatous form.^[4] The *cysticercus* larva (after

embedding itself in the cerebral parenchyma) undergoes four stages of evolution: vesicular, vesicular colloid, granular nodular, and nodular calcified.^[5] However, such evolution of the cysts is not appreciated in racemose NCC. Racemose NCC is rarely seen in India.^[6] We report a case of racemose NCC presenting as dementia.

Case Report

A 62-year-old lady presented with headache, slowness in activities of daily living and progressive memory disturbances of 8 months duration. For the last 6 months, she also had gait disturbance with slowness in activities of daily living and urinary incontinence. Her general examination was normal. Systemic examination was unremarkable. She had impairment of higher mental functions with Mini Mental Score Examination (MMSE) of 10/30. Cognitive domains severely affected were executive function, recent memory, visuospatial skills and calculation. Cranial nerves were normal and fundus examination did not show any papilledema. Motor examination revealed increased tone in all four limbs with cogwheel rigidity. Power was normal and deep tendon reflexes were brisk. Plantar reflex was flexor bilaterally. Sensory examination was normal. She had short shuffling gait. The hemogram was normal and biochemical parameters were within normal limits. She underwent magnetic resonance imaging (MRI) of the brain, which showed multiple variable-sized solitary and conglomerate cystic lesions in the right sylvian fissure, basal cisterns, sulcal spaces and third ventricle, which were

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hypointense on T1-weighted and fluid-attenuated inversion recovery (FLAIR) sequences and hyperintense on T2-weighted sequences [Figure 1]. No scolex was seen within the cysts. The anterior part of right lateral ventricle was pushed medially by conglomerate cystic lesions in the right sylvian fissure, but there was no midline shift. There was obstructive hydrocephalus with dilatation of lateral ventricles and third ventricle. Fourth ventricle was not dilated and cause of hydrocephalus was cyst in the third ventricle. Contrast-enhanced images showed patchy enhancement of cyst walls and septae [Figure 2]. There was no perilesional edema or any evidence of arachnoiditis. Diagnosis of racemose NCC was kept. She was given high-dose steroids and underwent ventriculoperitoneal shunting. NCC serology (ELISA for IgG antibodies) in serum and cerebrospinal fluid (CSF) was positive with titers of 1:1600 and 1:80, respectively. Post shunting, she had significant improvement in her cognitive status, with MMSE improving to 22/30 at the time of discharge 6 days post shunting. Her gait also improved and she became continent. She was discharged on oral steroids. Cysticidal therapy was withheld due to concerns regarding provoking acute brain inflammation and ependymitis from the death of cysts.

Discussion

Tenia solium is the most common cause of NCC, and its larval form *Cysticercus cellulose* consists of a bladder with one scolex. Cellulose cysts are usually located intra-axially in the brain tissue. The racemose form of cysticercosis is the less common presentation of NCC. As compared with *Cysticercus cellulose*, it is characterized by larger size, absence of scolex and extra-axial location. The cysts of racemose NCC often form grape-like clusters of multiple cysts that occur in subarachnoid/cisternal areas in and around the brain, such as the suprasellar, sylvian and quadrigeminal cisterns. According to some authors, racemose cysticercus may actually be due to a different variety of cestode. It could either be a sterile coenurus of *Taenia*

multiceps or, *Taenia serialis*, or an aberrant cysticercus of *Taenia solium*.^[7]

The most frequent clinical presentations of NCC are epilepsy, focal neurologic deficits and headache.^[4] However, as compared with parenchymatous NCC, which most commonly presents as seizures, racemose NCC due to its extra-axial location presents with raised intracranial pressure and meningitis. The subarachnoid space cysts can give rise to focal mass effect or to an inflammatory response manifested as basilar meningitis. Communicating hydrocephalus and signs of elevated intracranial pressure, such as nausea, vomiting, headache, and papilledema, may follow. Cranial neuropathy or vasculitis may result from the inflammatory changes, leading to small- or large-vessel infarcts.^[4,8]

Cognitive disturbances have been reported to occur in 66-87.5% of patients with NCC, and severe cognitive decline or dementia has been described in 12.5-15.6% of the patients with NCC.^[9-11] In one of the studies, the most frequently affected cognitive domains were executive functions, verbal memory, and language and visuospatial skills.^[9] Cognitive disturbances in NCC can be the result of multiple factors acting alone or in combination. Cognitive disturbances may be due to the mechanisms directly related to NCC or secondary to epilepsy caused by NCC as well as antiepileptic therapy. However, dementia as a sole presenting manifestation of NCC, as seen in the present case, is rare. Apart from raised intracranial pressure, other proposed primary mechanisms for cognitive decline in NCC include interaction among number of NCC, their localization, different evolutionary phases, the local production of inflammatory cytokines and host's immune response.^[9] The outcome of dementia in patients with NCC seems favorable in most cases. In one longitudinal follow-up study, complete or partial recovery was seen at 6 months follow-up in most cases (78.5%) treated with standard pharmacological therapy.^[10] The degree of reversibility of dementia seems to depend on predominant underlying mechanism with excellent outcome in patients like in this report, where raised intracranial

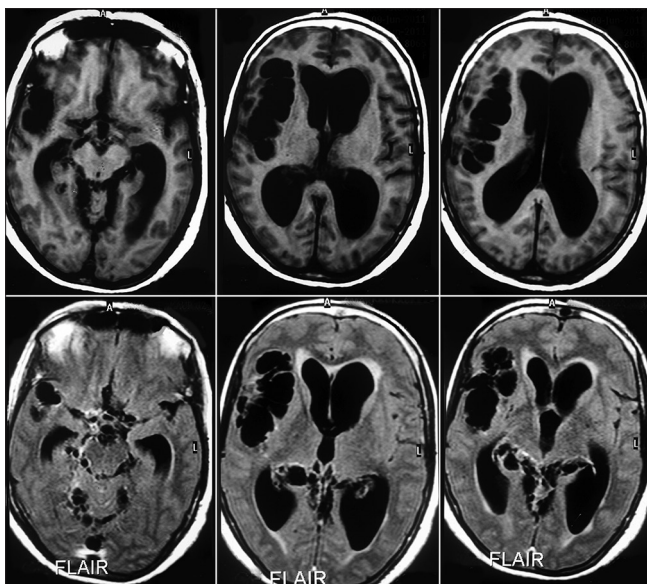


Figure 1: MRI brain T1 and FLAIR axial images showing conglomerate cystic lesions in right sylvian fissure and basal cisterns with hydrocephalus

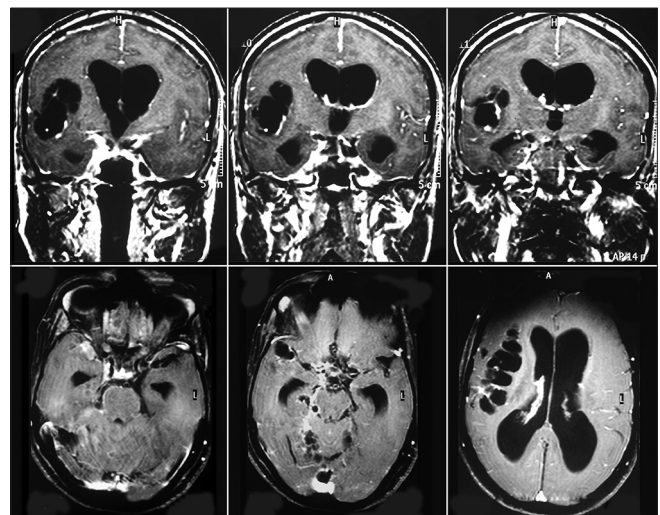


Figure 2: CE-MRI brain T1 coronal and axial images showing conglomerate cystic lesions in right sylvian fissure, third ventricle and basal cisterns with hydrocephalus and patchy cyst wall enhancement

pressure is the cause as compared with patients with multiple parenchymal parasitic and vascular lesions.

Accurate diagnosis is based on the combination of clinical, epidemiologic, radiographic and immunologic information. Immunologic studies used include detection of anticysticercal antibodies to *T. solium* antigens in the serum and CSF by enzyme-linked immunosorbent assay (ELISA) and enzyme-linked immunoelectrotransfer blot (EITB), HP10 antigen detection by ELISA and detection of parasite DNA by polymerase chain reaction (PCR). Of these, antibody ELISA, which was used in the present case, and EITB are the two most useful tests with CSF sensitivity equivalent to PCR (> 95%) with much better specificity (90% vs. 80%, respectively) in subarachnoid/cisternal NCC.^[12] For antigen ELISA test, CSF sensitivity of up to 86% and specificity of 100% has been reported in subarachnoid NCC.^[12] MRI is the imaging study of choice because it is more sensitive than CT for all phases of the disease process, with the exception of end-stage calcified disease.^[2] Biopsy of brain may still be required if diagnosis remains uncertain.

The treatment options for racemose NCC include medical therapy (cysticidal drugs and steroids), CSF diversion and surgical resection. Some advocate surgical resection as the best treatment for racemose NCC, while others recommend an initial trial of medical treatment in clinically stable patients.^[8,13,14] However, it may not be possible to remove all the cysts, and incomplete removal as well as cysticidal therapy alone may be associated with severe ependymitis and acute neurological deterioration; therefore, CSF diversion procedures without removal of cysts may be the best option in many of these patients. Dementia patients with NCC by large seem to have favorable outcome with appropriate therapy; therefore, NCC should always be considered in the differential diagnosis of dementia, especially in endemic zones.

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