Cysticercal Encephalitis: an Unusual Differential in the List of Encephalitis

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ABSTRACT

Introduction: Neurocysticercosis (NCC) is the most common infestation of Central Nervous System with majority of cases presenting with seizures.

Case report: Here we report a case of NCC in a young boy who presented as encephalitis which is a very rare initial presentation of the disease. MRI brain showed characteristic "starry sky pattern" with eccentric mural nodule specific for NCC. Cysticercal encephalitic is a very rare presentation of NCC which itself is an unusual cause of encephalitis.

Conclusion: It should be included in differential diagnosis of acute encephalitic cases apart from common causes especially in endemic countries like India. Early diagnosis and appropriate therapy can result in better outcome in this rare but commonly fatal neurological disorder.

Keywords: Neurocysticercosis, Cysticercal encephalitis

INTRODUCTION

Neurocysticercosis (NCC) is the most prevalent parasitic infestation in India. Central nervous system (CNS) involvement is seen in 60–90% of all infested patients with wide variations in clinical manifestations.[1] 70 to 90% of cases present with seizures while other common manifestations are headache, neurological deficit, hydrocephalus and raised intracranial pressure.[2,3] Seizure can occur both with degenerating cysts or calcified lesions. Rare neurological manifestations are spinal cysticercosis, progressive cord compression, ophthalmic cysticercosis, migraine and altered mental state.[3] Cysticercal encephalitis, is a very unusual presentation of NCC which occurs when there is diffuse cerebral edema along with multiple parenchymal cyst.[4] These patients are at risk of severe neurological sequelae because of brisk inflammatory response secondary to massive infection. Both MRI and CT can show the presence of an eccentric mural nodule which is characteristic of NCC. Seizures, being the most common presentation are usually controlled with standard antiepileptic drugs. Cysticidal therapy usually hastens radiological resolution of cysts but sometime associated with an exacerbation of neurological symptoms and death in some patients who have multiple cysts. Cysticercal encephalitis as manifestation of disseminated neurocysticercosis is very rare and only 0.3% of all cases of NCC have been reported in one large series of Indian children.[5] We present a case of acute cysticercal encephalitis in a 12 year old boy with characteristic imaging findings.

CASE REPORT

A 12 year old boy presented with complaints of progressive deterioration in consciousness since two days and single episode of seizure like activity one day back. There was history of on and off fever and headache for last 5 months with no history of head trauma, ear discharge, vomiting, jaundice or any history of tuberculosis contact. On examination child was afebrile with glasgow coma scale (GCS) of 4/15 while the vitals were pulse-110/min, RR-24/min, BP-130/70mmHg respectively. In CNS examination only positive finding was extensor plantar response and rest were unremarkable as GCS was very low. Chest examination revealed bilateral crepts while cardiovascular system and other systemic evaluation was within normal limit. In view of altered sensorium a differential diagnosis of encephalitis, encephalopathy or any space occupying lesions was made and investigated accordingly. Laboratory investigations showed Hb-10.9gm/dl, TLC-12x10^9 cells/L DLC-P90, L10, E10, platelet count- 650 ×10^9/L, Blood sugar- 110mg/dL, Blood urea-15mg/dL, S.creatinine – 0.3mg/dL, S.sodium-137mmol/L and S.potassium-4.8mmol/L. Malaria serology and mantoux test were negative and chest X ray showed mild perihilar opacities. The protein, sugar and ADA levels in CSF were 90mg/dL, 50mg/dL, and 8U/L respectively while total cells were 50 (P 45, L 5). MRI brain was suggestive of disseminated cerebral cysticercosis in various stages of evolution in bilateral cerebral hemisphere giving it the characteristic "starry sky pattern" (figure-1). Enhancing scolex was also noted in many of the cystic lesion with flair images revealing diffuse cerebral edema (figure-2). Child was managed initially with anti-convulsant along with antibiotics for prevention of secondary lung infection. Dexamethasone was given interavenously for 5 days followed by oral prednisolone over next 4 weeks with tapering. Albendazole was not given initially as it is contraindicated in such cases but after one week of symptomatic improvement, it was given for 8 days in a dose of 15mg/kg. After seven days of admission, patient regained consciousness but remained highly irritable. He had variable paresis in all four limbs for which he was put on regular physiotherapy and later on discharged. On follow up after 6 weeks, child's behaviour improved remarkably but weakness and abnormal gait resolved only after 6 months. Repeat CT scan after 6 months showed only few ring enhancing lesions as compared to earlier scan (figure-3).

DISCUSSION

Neurocysticysticercosis is the most common parasitic disease of the CNS caused by ingestion of foods contaminated with Taenia solium. The cysticercal larva after reaching the brain invokes only minimal inflammatory reaction and forms cyst which

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later degenerates with thickening of wall and opacification which are easily picked up by neuroimaging as contrast-enhancing ring like lesions. Cysticerci may be located in brain parenchyma, subarachnoid space, ventricular system or spinal cord causing pathological changes that are responsible for the pleomorphism of neurocysticercosis.[6] Though seizures are most common initial presentation of NCC but in the present case child presented in a state of altered sensorium for past two days with headache and insignificant history of single seizure like activity. Cystecercal encephalitic is a severe and rare form of neurocysticercosis. Besides that only few cases in children have been reported till yet.[7] It is characterized by altered sensorium, seizures, diminution of visual acuity, headache, vomiting, and papilledema as in the present case.[4] This occurs when there is massive cysticerci infection of the brain parenchyma and the host's immune system actively reacts against the parasites. Although enhancing lesions are typical of neurocysticercosis but they can be noted with tuberculomas, brain abscess and tumours. Accurate diagnosis of NCC is possible after interpretation of clinical data together with findings of neuroimaging studies and results of immunological tests. Despite the advances in neuroimaging and immune diagnostic tests, the diagnosis of neurocysticercosis is a challenge in many patients. Clinical manifestations are nonspecific, neuroimaging findings are often not pathognomonic, and serologic tests are faced with problems related to relatively poor specificity and sensitivity.[8] So a set of diagnostic criteria based on clinical, radiological, immunological and epidemiological data are used to diagnose patient with suspected NCC. This includes four categories of criteria- absolute, major, minor and epidemiological (Table-1).[8] Any suspected patient can be diagnosed as definitive case if there is presence of one absolute criteria and two major plus

Revised Diagnostic Criteria for Neuro-cysticercosis	
Absolute	
1.	Histological demonstration of the parasite from biopsy of brain or spinal cord lesion
2.	Cystic lesions with scolex on CT or MRI
3.	Direct visualization of subretinal parasite by fundoscopy
4.	Spontaneous resolution of small single enhancing lesions
Major	
1.	Lesions highly suggestive of NCC on neuroimaging
2.	Positive serum immuno blot for detection of anti-cysticercal antibodies
3.	Resolution of cysts after antiparasitic therapy
Minor	
1.	Lesions compatible with NCC on neuroimaging
2.	Clinical manifestations suggestive of NCC
3.	Positive CSF-ELISA for detection of anticysticereal antibodies or cysticercal antigens
4.	Cysticercosis outside the CNS
Epidemilologic	
1.	Individual coming from or living in an endemic area
2.	Evidence of household contact with Taenia solium infection.
3.	History of frequent travel to disease-endemic areas
Definitive	
1.	Presence of one absolute criterion or two major plus one minor and one epidemiologic criteria

Table-1: Criteria for NCC diagnosis

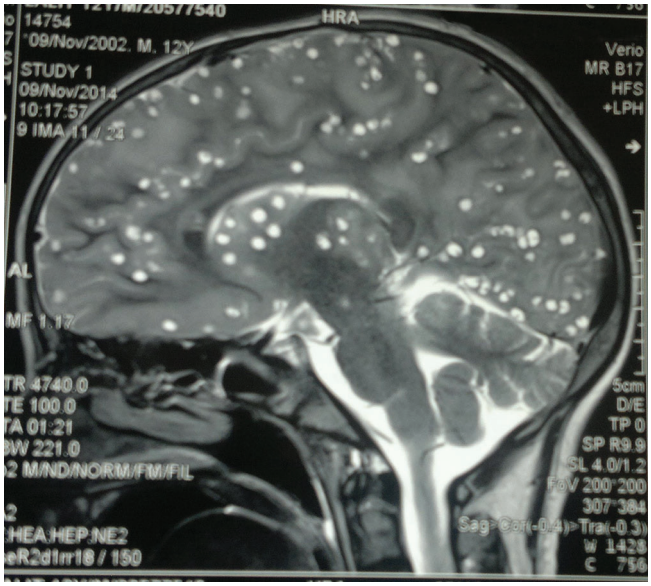

Figure-1: MRI brain showing multiple solid and cystic lesion appearing hyperintense on T2 W image giving the characteristic "Starry sky appearance".

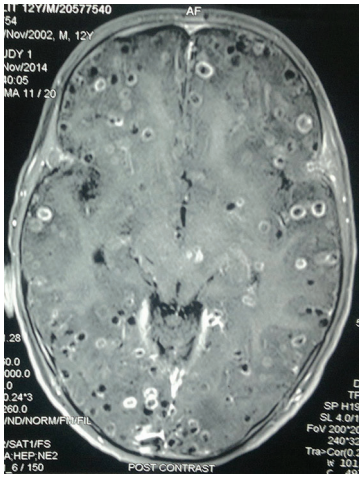

Figure-2: MRI brain showing multiple ring enhancing lesion with eccentric mural nodule in few of them

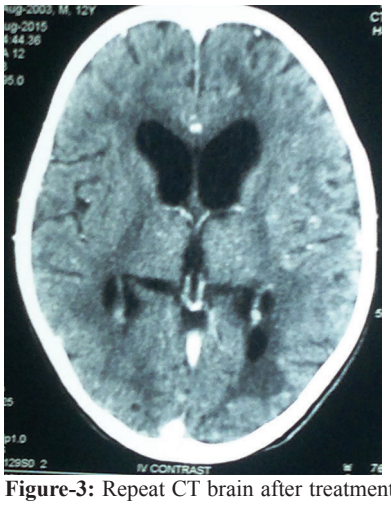

Figure-3: Repeat CT brain after treatment showing only few remnant calcified lesion.

one minor or one epidemiological criteria.[8] This case full fills criteria for definitive diagnosis. The current consensus is that patient with multiple parenchymal cysticerci should be treated with course of albendazole 15 mg/kg/day for 8 days along with simultaneous course of corticosteroids apart from anti-epileptics.[9] But in this case albendazole was given after one week, in view of their potential to exacerbate host inflammatory response. Patient with numerous cysticerci may require repeated courses of therapy and even surgery.[10]

CONCLUSION

Accurate diagnosis and early appropriate therapy can result in better outcome in such rare but commonly fatal neurological disorder.

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