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Dementia as the presenting manifestation of neurocysticercosis: A report of two patients

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Abstract

Neurocysticercosis (NCC) is the commonest parasitic disease of central nervous system and an endemic problem in India. Its clinical manifestations are varied, non specific and pleomorphic, depending on number and topography of lesions. We report two patients of NCC, who presented only with dementia. Dementia improved in both of them, by using steroids as cerebral decongestant. Since NCC mimic large number of neurological disorders, it is important that clinicians should be familiar with these rare presentations to avoid delay in diagnosis and management. This report further suggests the utility of decongestive therapy in multiple NCC. It also heralds that dementia can sometimes be a reversible syndrome, if diagnosis and its etiology is established early and correctly.

INTRODUCTION

Neurocysticercosis (NCC) is the most common helminthic infestation of the nervous system in developing countries including India.[1,2] Clinical manifestations are varied, non specific and pleomorphic, depending on number and topography of lesions. Besides seizures, which are commonest, other clinical features include raised intracranial pressure, syndromes of cerebellopontine angle lesion, dementia and variety of strokes.[3-5] We report two patients of NCC presenting with dementia as the only predominant manifestation.

CASE REPORT 1

A 45-year-old male shopkeeper, non alcoholic, educated up to college, presented with bouts of severe headache, vomiting and gradually progressive decline in cognitive functions of 3 months duration. His decline in cognition was hampering his daily routine activities. He forgot taking meals, remained unconcerned for personal care and misplaced things easily. He made gross errors in calculations and collecting payment from customers. There was difficulty in concentrating on his business, resulting in heavy financial loss. He could not easily recollect the names of his close relatives. All these behavioral changes made him very irritable, depressed and anxious. The wife got alarmed when he forgot his way home and was brought home by his neighbors, who informed that he was behaving like a drunk, and had left the shop without closing it properly. His wife consulted a psychiatrist who referred him to our tertiary care centre.

On examination he was unkempt and clothes were untidy. The general physical and systemic examination was normal. Although he was conscious and alert, he was resisting evaluation. He declined that he was ill and required help. There was significant cognitive decline and dementia as his Folstein's Mini Mental Status Examination (MMSE) score was 20. The speech was normal. There were no focal neurological deficits and the cranial nerves were intact. The fundus examination was normal. Hemogram, hepatorenal function and electrolytes were normal. Thyroid function tests and vitamin B-12 levels were within normal range. Electroencephalogram (EEG) was normal. Magnetic resonance imaging (MRI) revealed multiple cysts involving the whole of cerebral cortex (Figure 1). Some of the cysts had prominent scolex suggestive of NCC. There was no evidence of ventricular dilatation or intracerebral edema. A short course (10 days) of prednisolone 1 mg/kg/day was given initially, followed by acetazolamide 250 mg thrice a day for 3 months. Headache and vomiting subsided completely over 10 days. The dementia improved (MMSE score elevated to 24) over initial 2 weeks, but later remained stationary for other one year. The MMSE score returned to normal by the end of 2 years (Table1) when he was able to look after himself and his occupation independently.

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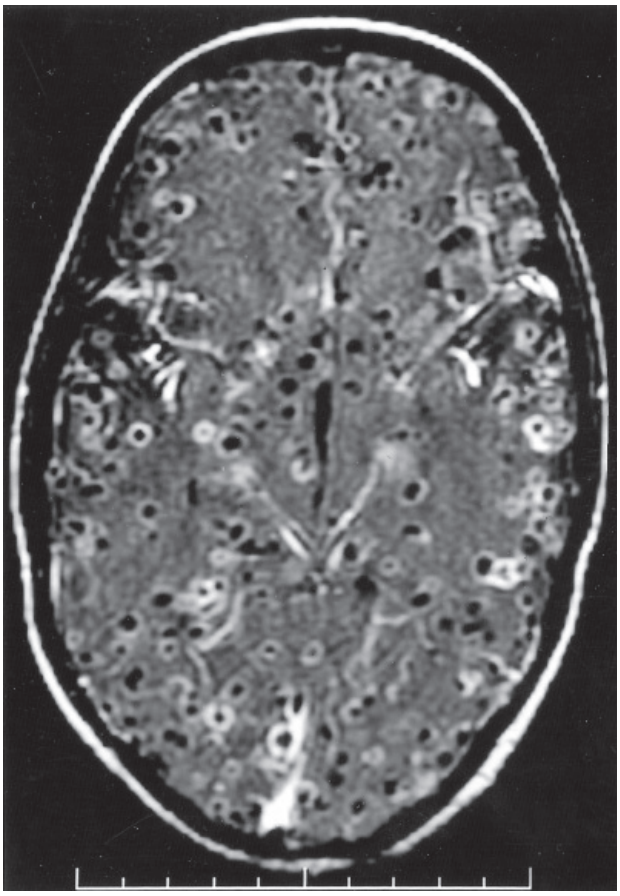

Figure 1. T_1 weighted contrast MRI sequence revealing multiple ring enhancing lesions, diffusely scattered throughout both the cerebral hemispheres in cortical and sub cortical white matter with eccentric scolex.

CASE REPORT 2

A 42-year-old graduate, businessman by profession, presented with progressive headache with nausea and vomiting of 5 months and forgetfulness of 3 months duration. He made gross errors in day to day activities, like forgetting whether he has consumed meals or not, important tax files, and delay in recognizing relatives. The relatives got alarmed when he made gross errors in calculation related to his business. He started behaving like an alcoholic, whereas he was a teetotaler. He took lots of time in recollecting whereabouts of his belongings. Initially he helped his children in their school assignments, but then he started making constant errors and blunders. A psychiatric consultation was sought after which he was referred for neurological consultation.

The general and systemic examination was normal. Except for a decreased score in MMSE (18), there was no neurological deficit. Investigations for complete hemogram, renal and liver function tests, thyroid function tests, Vitamin B12 level and electrolytes were within normal limits. EEG was normal. Cranial MRI showed diffused and widespread cysts with prominent scolex (Figure 2). The patient was treated with steroids initially, followed by acetazolamide. His MMSE score improved to 26 and symptoms of headache and vomiting disappeared within 2 weeks. (Table 1) He could perform his routine

Table 1: Follow up MMSE Score of both patients with neurocysticercosis

	Base line	2 weeks	6 months	12 months	18 months	24 months
Case 1	20	24	25	24	29	30
Case 2	18	26	26	25	28	30

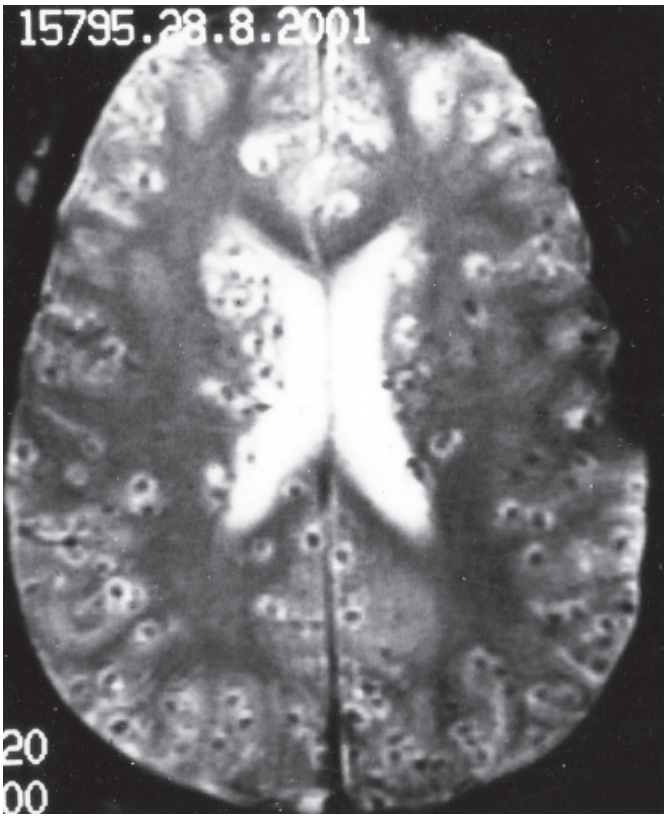

Figure 2. T_2 weighted image revealing multiple ring enhancing lesions with prominent hypointensity in center surrounded by hyper intensity at the periphery with eccentric scolex scattered throughout grey and white matter. They are also present in the lateral ventricles and periventricular regions.

activities and look after his family and business independently

DISCUSSION

The diagnosis of NCC in both our patients was confirmed on the basis of clinical, radiological and epidemiological criterias[3,6], after excluding other etiologies mimicking NCC, like tuberculomas, fungal granulomas or toxoplasmosis.

The interesting features in both patients were dementia as the presenting symptom and absence of seizure or any focal neurological deficit. Though infectious diseases represent an important cause of behavioral changes including dementia, there are only few isolated reports of NCC presenting with cognitive disturbances as the sole manifestation.[7-10]

Despite extensive involvement and multiple NCC, there was no edema or gliosis apparent from conventional imaging in both the cases. There are numerous MR imaging techniques like diffusion-weighted (DW) imaging with apparent diffusion coefficient (ADC), FLAIR (Fluid Attenuation Inversion Recovery) and magnetization transfer magnetic resonance imaging (MT sequence), which are more specific for evaluation and differentiation of perilesional edema and reactive gliosis in NCC. However, due to technical reasons, they were not performed.

Both the cases were managed with steroids and decongestive therapy. No cysticidal drug was given since it may sometimes be detrimental, especially in patient like ours.[11] There were multiple lesions and sudden anaphylactic reaction and deaths are reported to be more common in multiple NCC, when treated with cysticidal drug.[12] The Cochrane Database review on drugs for treating NCC concludes, that there is insufficient evidence to assess whether cysticidal therapy in NCC is associated with beneficial effects.[13] Parenchymal NCC can resolve on imaging studies without being treated with antiparasitic drugs.[14,15] Acute, severe brain inflammation result from cysticidal drugs and their use is reported to be unnecessary because parenchymal brain cysticercosis follow a benign course and cysts will degenerate and heal by natural evolution of the disease.[7,16-19] In

both the cases there were multiple ring enhancing lesions suggestive of heavy infection burden with degenerating cysts, so high-dose steroids and osmotic diuretics were started and antiparasitic treatment was deferred.[20]

There is conflicting evidence to support use of steroids as the primary treatment in NCC. However in subarachnoid cysts, chronic meningitic form or in case of multiple cysts, steroid should be given with cysticidal drugs.

Both our patients became asymptomatic and showed resolution of dementia within 2-3 weeks. They were followed up for another 2 years using MMSE to help evaluate the mental function. The radiological follow up is equally important as the perilesional edema may not always subside with treatment, and may at times be present around apparently calcified foci. Perilesional gliosis may also persists predicting long term AED requirement.[21]

Dementia observed in patients with NCC is possibly due to the combined effect of multiple factors, viz; multiple parasitic lesions, vascular involvement, disruption of frontal-parietal-temporal networks, raised intracranial tension[22] and inflammatory injury of brain parenchyma.[23] Dementia in NCC is reversible as was evident in both patients, similar to earlier reports.[22] An interesting observation in both patients was that even in the absence of clinical or radiological changes of raised intracranial tension, decongestant therapy led to reversal of dementia. Other potential mechanism, postulated for cognitive decline is inflammatory reaction that accompanies the death of parasite with edema and dysfunction of perilesional tissue or mass effect[24,25] which may explain the cognitive impairment in these patients. Earlier reports have stated that patients with old age, lower educational level, increased number of parasitic lesions and involvement of the frontal, parietal and temporal lobes are more likely to develop dementia due to NCC.[22]

This case report highlights the protean manifestations of NCC and also underscores the importance of early diagnosis of NCC in patients with dementia from endemic areas. It is important to be aware of this rare presentation to avoid erroneous diagnosis and delay in management. This report further suggests the utility of decongestive therapy in multiple NCC. It also shows that dementia can sometimes be a reversible syndrome, if diagnosis and its etiology is established early.

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