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Multiple neurocysticercosis (more than 400 lesions) with intraventricular involvement

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Abstract. Neurocysticercosis (NCC) is the most common central nervous system parasitic disease worldwide, but cysticercal meningitis and intraventricular lesions are relatively rare, especially in Indian patients. Disseminated NCC with numerous cysts may give rise to the so-called "starry-sky" appearance on neuroimaging studies. Magnetic resonance imaging allows optimal identification of scolex and visualization of extraparenchymal cysts. We herein report a girl with multiple NCC (more than 400 lesions) with intraventricular involvement, but without focal neurological deficit on examination.

Keywords: Multiple neurocysticercosis, intraventricular cyst, "starry-sky" appearance

1. Introduction

Neurocysticercosis (NCC) is the most common parasitic infestation of the central nervous system worldwide. In India, the parenchymal variety of NCC is the most common type [1], while intraventricular cysticercosis is uncommon. In one study of 374 consecutive pediatric NCC patients, only 0.3% had the intraventricular variety [1]. Most children (> 80%) present with partial seizures, although the presentation varies depending on the stage and location of the cysts in the nervous system. About a third of affected patients present with headache and vomiting. Status epilepticus has been reported in 1.7% to 32% of cases [2]. Diagnosis is made by either computed tomography (CT) or magnetic resonance imaging (MRI). Single enhancing lesions are the most common finding. Visualization of a scolex confirms the diagnosis. Some patients have multiple cysts with a characteristic "starry-sky" appearance on neuroimaging. Management involves use of anticonvulsants for seizures and steroids for cerebral edema. The use of cysticidal therapy continues to be debated. Controlled studies have shown that cysticidal therapy improves and speeds resolution of lesions identified on CT-scanning. Improvement of long-term seizure control has not yet been proven. Children with single lesions have a good outcome and low seizure recurrence rate. Children with multiple lesions, particularly those with disseminated NCC and calcifications, have frequent seizure recurrences. Extraparenchymal NCC is rare in children as compared to adults [3].

2. Case report

A 5-year-old girl with normal physical and mental development came to our hospital in convulsive status epilepticus with loss of consciousness. She had a history of two episodes of non-projectile vomiting preceding the convulsions. There was no history of fever, dizziness, vision changes, headache or altered sensorium before the onset of seizures. Past history revealed that 20 days prior to admission, the child had had multiple episodes of vomiting. A CT scan was done in

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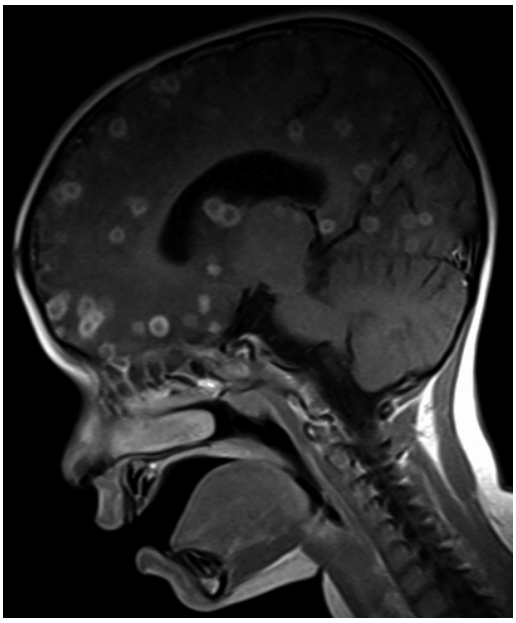

Fig. 1. Magnetic resonance imaging of brain axial cuts showing multiple neurocysticercosis in the brain parenchyma, "starry-sky" appearance.

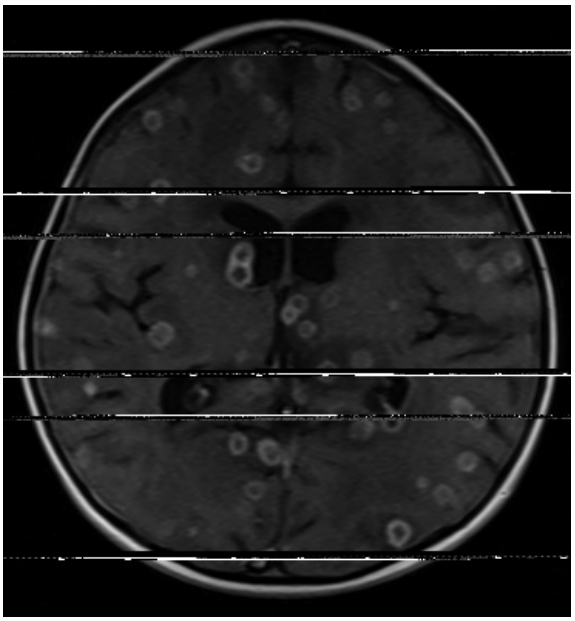

Fig. 2. Magnetic resonance imaging of brain axial cuts showing intraventricular cysts.

a private hospital and led to a diagnosis of multiple NCC. Intraventricular lesions were not noted on the CT scan. The child was started on enteral therapy with albendazole, which was taken for 15 days, and steroids (dexamethasone), which were taken for 5 days.

On admission the child was actively convulsing. Her seizures were controlled with parenteral phenytoin (40 mg/kg) and phenobarbital (20 mg/kg), after which maintenance doses of the same medications were prescribed. Primary assessment revealed that the child was afebrile, heart rate was 135 beats/min with good central and peripheral pulses, and blood pressure was 86/60 mmHg. There were no signs of meningeal irritation. Fundoscopy revealed bilateral papilledema. MRI of the brain performed in our hospital revealed multiple NCC (more than 400 lesions) with intraventricular involvement and a characteristic "starry-sky" appearance (Fig. 1). Lumbar puncture was initially delayed due to the papilledema, but was performed the next day. The opening pressure was raised, but cerebrospinal fluid (CSF) analysis was normal. A chest x-ray was normal. The Mantoux test was negative. Anti-edema measures were started with parenteral mannitol (0.5 g/kg/day) and dexamethasone (0.15 mg/kg/day).

There were no further convulsions in the hospital. The child had postictal drowsiness lasting for 1 hr, after which her condition gradually improved. The child became fully conscious after 24 hrs. She didn't have any focal neurological deficits at any time during the hospital stay. Gradually parenteral phenytoin and phenobarbital were tapered and enteral therapy was initiated with carbamazepine (10 mg/kg/day). Mannitol was stopped after 3 days, while dexamethasone was continued for 5 days. She was discharged on oral carbamazepine and is being followed regularly.

3. Discussion

NCC is a major cause of neurological disease worldwide [4,5]. It is an important cause of epilepsy in the tropics [6]. NCC is caused by infestation of the central nervous system with encysted larvae of *Taenia solium*. Infection is acquired through the fecal-oral route when humans consume raw vegetables contaminated with *T. solium* eggs or food prepared by carriers of tapeworms. NCC is generally classified into (a) parenchymal and (b) extraparenchymal type, whereby the latter includes ventricular, cisternal, ophthalmic or spinal forms. Most children present with single degenerating parenchymal cysts, some with multiple cysts. It is not well understood as to why some cases have single and others have multiple cysts. Immunological differences may possibly account for this; defective functions of neutrophils and T lymphocytes have been reported in patients with multiple lesions only and not in those with single le-

sions [7]. Ventricular and subarachnoid NCC generally occur together and present as basilar arachnoiditis, obstructive hydrocephalus or chronic meningitis [2]. In most cases the lesions are single and < 20 mm in size – termed as single, small, enhancing lesions on computed tomography [8]. Some children may have multiple lesions. Disseminated NCC with numerous cysts may give rise to the characteristic, so-called "starry-sky" appearance. MRI is better than CT-scanning at identification of scolex and visualization of extraparenchymal cysts (Fig. 2) [9].

CT scanning has a sensitivity and specificity of over 95% for the diagnosis of NCC [10]. However, the sensitivity of CT is much lower for ventricular or cisternal forms of the disease. MRI is the most accurate technique to assess the degree of infection, the location and the evolutionary stage of the parasites. It provides excellent visualization of perilesional edema, of the degenerative changes of the parasite, and of small cysts or those located in the ventricles, brainstem, cerebellum, base of the brain, eye and spine [11]. CT is more sensitive for the detection of calcifications. The main disadvantages of MRI are its high cost and limited availability. Thus, in our setup, CT scan is usually the first investigation, while MRI imaging is reserved for patients with inconclusive CT findings. Because of the high incidence of small enhancing computerized tomographic lesions in our country, a CT scan is indicated after a first focal seizure.

There are four stages of development of a parenchymal larval cyst: vesicular, colloidal, nodular and calcified. The vesicular cyst is viable. The scolex exists as an eccentric nodule within the cyst, and there is little or no enhancement due to a minimal host immune response. As the scolex dies after cysticidal treatment or an effective immune response, the transparent vesicular fluid is replaced by a viscous and turbid fluid, which is readily identifiable on MRI. The fluid migrates from the degenerating cyst into the surrounding parenchyma and incites a robust immune response characterized by strong enhancement on contrast CT scans or MRI. This is the colloidal cyst, which has contrast enhancement, but lacks a well-defined scolex. As the cyst further degenerates, it develops into a nodular cyst, which still shows some contrast enhancement. Finally the degenerated cyst calcifies and is recognized as a punctuate calcification on CT scan.

The "starry-sky" appearance is not just a function of a dead calcified scolex, but it appears even in other stages of NCC (i.e., the vesicular, colloidal and nodular stages). In disseminated or miliary NCC, there are multiple cysts in varying stages of development (i.e. living, dying and calcified cysts). On a CT-scan, cysts appear as single or multiple rounded lesions of variable size and low density, with a small, hyperdense, eccentric mural nodule representing the scolex. This gives a "starry-sky" effect in the parenchyma. Ring enhancement occurs with an inflammatory reaction or a granuloma formation. On brain MRI, well-defined vesicular cysts with eccentric scolices appear as a "hole-with-a-dot" sign. Cysts in the colloid-vesicular stage can show perilesional edema and ring enhancement. This also gives rise to "starry-sky" patterns because of multiple cysts in the brain parenchyma [12].

Intraventricular and subarachnoid (cisternal) NCC generally occur together and present as basilar arachnoiditis, obstructive hydrocephalous or chronic meningitis. Some subarachnoid cysts may enlarge without scolices. They may become racemose and cause mass effect [3]. The oncosphere reaches the ventricular cavity via the choroid plexus. The parasite migrates through the ventricular system occluding CSF communication corridors, causing acute episodes of ventriculomegaly with sudden death, or mass effect with focal compression.

MRI is the most accurate technique to assess degree of infection, location, and evolutionary stage of the parasite. It is possible to differentiate the various cyst stages on MRI. The intensity of fluid in live cysts is similar to CSF. The scolex appears as a mural nodule of high signal intensity on T1-weighted MRI and low signal intensity on T2-weighted MRI (like a "hole with a dot", or a "pea in a pod"). There is no perilesional edema at this stage. In a degenerated cyst, the fluid becomes turbid (colloid vesicular stage), appearing as a high intensity signal on a T1-weighted MRI. In the granulonodular stage, ring enhancement occurs with gadolinium injection, which is isointense on T1-weighted and hypointense on T2-weighted images. There is variable perilesional edema. The racemose type of cyst is a large lobulated cyst without a scolex, whereas the cellulose type contains a scolex inside a vesicle. Intraventricular cysts are well delineated with a small metacystode inside the cyst. Abnormal enhancement after gadolinium suggests ependymitis or ventricular entrapment [12].

Treatment of intraventricular cysticercosis is a contentious issue. Medical management has fallen out of favor because of poor cysticidal CSF penetration, delay in response and, lysis of the cyst leading to ependymitis with resultant acute complications and chronic sequelae [13]. Placement of a ventricular shunt and medi-

cal therapy to prevent or treat hydrocephalus are usually required. Shunt dysfunction rate is also high (30-67%) [14]. Surgery eliminates the inflammatory nidus, potentially obviating the need for a complication-prone shunt. Open surgery was previously the only available modality, but now neuroendoscopy has become the treatment of choice [13,15–17].

Cysticidal therapy should not used in cases with markedly raised intracranial pressure (ICP), particularly in disseminated NCC, as sudden elevations of ICP may occur secondary to the host inflammatory response, and in ophthalmic NCC, as the host response may cause damage to the eye. Such cases should be treated with steroids alone. Cysticidal therapy is not indicated for calcified lesions as the parasite is already dead and buried. In view of the controversies involved, consensus guidelines for the treatment of NCC were published [17] and re-discussed [18].

In the present case, cysticidal therapy with albendazole had caused sudden elevation of ICP leading to all the above signs and symptoms. Also, the intraventricular cyst was not detected due to the poor sensitivity of CT-scans for this type of lesion. To conclude, cysticidal therapy should be avoided in cases of disseminated NCC and – although expensive – MRI is preferable to CT-scanning in all cases of suspected NCC with intraventricular involvement.

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