Intraventricular Neurocysticercosis (Racemose Form): A Rare Entity –A Case Report And Review Of Literature

V Velho, S Khan, M Sharma, D Palande

Citation

V Velho, S Khan, M Sharma, D Palande. *Intraventricular Neurocysticercosis (Racemose Form): A Rare Entity –A Case Report And Review Of Literature*. The Internet Journal of Neurosurgery. 2009 Volume 7 Number 1.

Abstract

Objective: Neurocysticercosis-racemose form is a rare parasitic infestation of the central nervous system. They can be misdiagnosed as hydatid cyst or intraventricular epidermoid thus presenting as a diagnostic dilemma.Settings: Grant Medical College & Sir J.J Group of Hospitals, byculla, Mumbai, Maharashtra, India. Methods: A 16 year old male presented with headache, vomiting, imbalance on walking and diminision of vision of short duration. CT scan and MRI scan with contrast was suggestive of a fourth ventricular lesion. The patient was operated upon with complete excision of the lesion. Results: The patient showed immediate neurological improvement with reduction in headache, vomiting and imbalance on walking. He has been following up regularly with gradual recovery. Conclusion: Neurocysticercosis- racemose form is a rare intraventricular lesion. Complete surgical excision followed by appropriate drug therapy should be given to achieve cure.

INTRODUCTION

Cysticercosis is a parasitic infection with CNS involvement in 60-90% of patients. It is the commonest parasitic disease of the central nervous system. CNS cysts are encountered in 4 types in neurocysticercosis: (1) meningeal (2) parenchymal (3) ventricular and (4) mixed. The ventricular system is the second most common site of neurocysticercosis. It is frequently caused by Cysticercus cellulosae, however Cysticercus racemosus can also infect the ventricular system The racemose form is characterized by proliferative lobulated cysts without a scolex. Although infrequent, this is the most serious manifestation of neurocysticercosis.

CASE REPORT

A 16 year old male presented with history of headache, vomiting, imbalance on walking and progressive diminution of vision of short duration. There was no history of fever and convulsions. A clinical examination revealed signs of raised intracranial pressure with bilateral cerebellar signs.

CT Scan of the brain was suggestive of a posterior fossa midline cystic mass with mass effect over the 4th ventricle with mild obstructive hydrocephalus.

MRI Brain (Fig. 1) revealed a well defined, 3.1 ×1.9 cm, non enhancing, lobulated, near CSF attenuation, mass lesion with restricted diffusion on DW images in the region of the 4th ventricle, with mass effect and moderate obstructive hydrocephalus and was suggestive of an epidermoid.

Suboccipital craniectomy with excision of the midline cystic SOL was done. The cyst was about 1.5-2cm deep to the surface of vermis. The cyst was thick walled, yellowish, non vascular, not adherent to the roof of 4th ventricle. It was removed in toto. On bisection, the interior of the cyst revealed multiple smaller vesicles with a bunch of grape appearance, raising the possibility of hydatid disease.

However the final neuropathology report (Fig.2) was Cysticercus cellulose

(Racemose form). Several interconnected bladders of various sizes were present. However there was no evidence of any scolex as seen in classical cysticercosis or a germinal membrane and laminated structure as seen in hydatid disease.

The patient was managed with a 4 week course of albendazole post operatively. The patient did well postoperatively, vision improved and ataxia was completely relieved.

Figure 1

Fig.1- MRI Scan shows a well defined, non enhancing, lobulated, mass lesion in the region of 4th ventricle, with mass effect and moderate obstructive hydrocephalus.

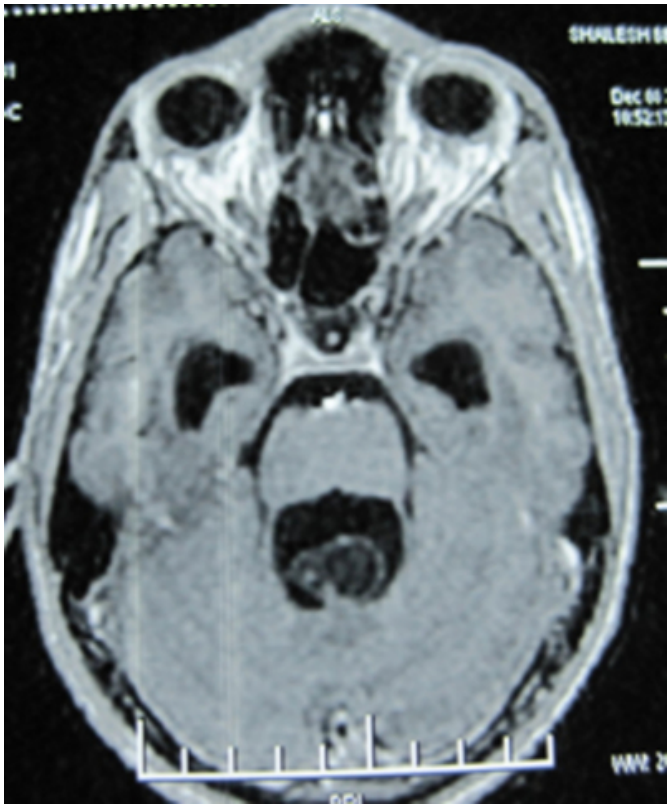
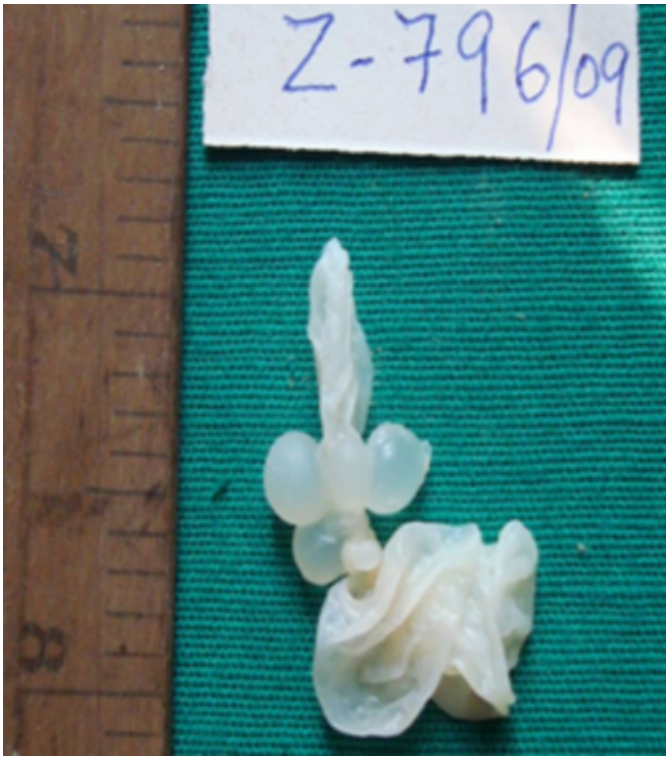


Figure 2

Fig.2- Histopathological examination showing Cysticercus cellulose (Racemose form) with several interconnected bladders of various sizes.



DISCUSSION

Cysticercosis of the CNS (neurocysticercosis) has a worldwide distribution. Neurocysticercosis is endemic in most of Central America, South America, and Asia.

It is caused by a human tapeworm, Taenia Solium which has a complex two-host life cycle. In humans, it can cause Taeniasis and cysticercosis. [(1)]

After entering the CNS the cysticerci are viable and elicit very little inflammation. It can remain like this for long time protected by the blood-brain barrier. After variable amount of time the cyst starts to degenerate resulting in inflammation. Cysts cause disease by acting as mass lesions, blocking CSF flow. Most of the symptoms are a direct result of the host inflammatory response due to cyst degeneration. The clinical manifestations depend on the number, location, size and host's immune response to the cysts. Neurocysticercosis can present as epilepsy, headache, paraparesis, psychiatric symptoms, stroke and dementia. While in the nervous system, the parasite goes through different stages of involution, which include the following [(2)]

Vesicular stage, a viable parasite with a mild inflammatory

reaction

Colloidal stage, a parasite with a scolex in the process of degeneration and a severe inflammatory reaction around it

Granular stage, a parasite with a degenerated scolex and astrocytic gliosis around the cyst

Calcified stage, a parasite transformed into a calcified nodule with intense gliosis around the cyst

Cysticercus is a liquid-filled vesicle with a 3-layer wall and scolex, although the scolex may not be found. The parasite can adopt 3 different presentations in the nervous system—cystic, racemose, and mixed form.

Cystic form refers to the presence of cysts anywhere in the brain; cysts are approximately 7 mm in diameter and may be single or multiple. Their most frequent locations are the leptomeninges and the cerebral cortex.

Racemose form refers to the presence of multiple cysts in the basal cisterns where the vesicles can have different sizes, and the cysts can be attached to the meninges. They are aberrant cysticerci with grapelike clusters of cysticerci.They do not have a scolex. The histopathological examination of the cyst removed from our patient revealed exactly similar findings.

Cyst wall grows in an irregular, branching and budding fashion with a diameter of several centimeters. Because of their location, they can produce hydrocephalus, which is caused by inflammation of meninges with subsequent fibrosis and obstruction. They have a poor prognosis, do not respond to drugs and must be surgically removed. [(3)]

CNS cysts are encountered in 4 types in neurocystcercosis: meningeal, parenchymal (solitary or multiple cysts), ventricular (usually solitary), and mixed.

Meningeal cysts form mostly in the basal meninges, sometimes causing stroke and hydrocephalus. Parenchymal cysts are usually found in the cerebral cortex, including the cortical-subcortical junction. The white matter is rarely involved. Ventricular cysts are seen in 15% of patients withneurocysticercosis; in 50% of cases, they are located in the fourth ventricle. They may cause intermittent hydrocephalus. In approximately 20% of cases, parenchymal cysts are found concomitantly with intraventricular cysts.

The ventricular system is the second most common site of neurocysticercosis[(4)]. It is frequently caused by Cysticercus cellulosae, however Cysticercus racemosus can also infect the ventricular system. The intraventricular form of the disease is found in more than 54% of patients with intracranial cysticercosis studied by MRI.

It most commonly affects the fourth ventricle (54%-64%), followed by the III ventricle (23%-27%), the lateral ventricles (11% - 14%) and Sylvius aqueduct (9%).

Intraventricular lesions are isodense to CSF on CT Scan.

MRI is sensitive in the diagnosis of active neurocysticercosis and may be useful in evaluating degenerative changes in the parasite. [(5)]

Intraventricular cysts are detected on MRI by mass effect, ventricular obstruction, and detection of a cyst rim and / or CSF flow void adjacent to the rim. Intensity of cysts that are viable is similar to that of CSF on both T1 and T2 weighted images.

Differential diagnosis includes other causes of intraventricular cystic lesions. Cystic hydatid disease almost always appears on CT/MRI as a single, large, spherical, and nonenhancing intracranial cyst. This is a very rare form of presentation of T. solium cysticercosis. The MRI of our patient showed a similar picture of a single, large, spherical, non enhancing fourth ventricular cyst, raising the suspicion of hydatid disease as well in addition to epidermoid. Also, the current assay of choice, immunoblot, does not cross-react with echinococcosis. Other condition that may resemble T. solium cysticercosis from the clinical and neuroimaging points of view is coenurosis, an extremely rare condition caused by the cestode Multiceps multiceps.

In India and other less developed countries the diagnosis of neurocysticercosis is frequently difficult because several other prevalent neurological disorders can present with a similar clinical and neuroimaging picture.

In our case, the initial radiological diagnosis was epidermoid cyst while intra-op it looked like a hydatid cyst due to the presence of multiple smaller cysts within. However the final histopathology report was racemose type of neurocysticercosis, which often grossly resembles hydatid cyst. The racemose variety which is found in the basal meninges and infrequently in the ventricles, is the most severe variety, does not respond well to medical treatment,

has a poor prognosis and surgical removal is often the only chance for cure.[5]

References

1. Gubbay AD, Brophy BP, Henley S, Sage M. Neurocysticercosis. J Clin Neurosci. Apr 1998; 5(2):203-7.
2. Martinez HR, Rangel-Guerra R, Arrendondo-Estrada JH, Marfil A, Onofre J. Medical and surgical treatment in neurocysticercosis: A MR study in 161 cases. J Neurol Sci 1995 May; 130(1): 25-34.
3. Am. J. Trop. Med. Hyg., 30(3), 1981, pp. 620-624
4. Mathews M, Paré L, Hasso A. Intraventricular cryptococcal cysts masquerading as racemose neurocysticercosis. Surg Neurol. Jun 2007; 67(6):647-9.
5. Rahalkar MD, Shetty DD, Kelkar AB, et al. The many faces of cysticercosis. Clin Radiol. Sep 2000; 55(9):668-74.

Author Information

Vernon Velho
Associate professor & Head of Dept, Department of Neurosurgery, Grant Medical College & Sir J.J Group of Hospitals

Shadma Khan
Resident, Department of Neurosurgery, Grant Medical College & Sir J.J Group of Hospitals

Mayur Sharma
Resident, Department of Neurosurgery, Grant Medical College & Sir J.J Group of Hospitals

Deepak A. Palande
Professor, Department of Neurosurgery, Grant Medical College & Sir J.J Group of Hospitals