Review

Surgical management of neurocysticercosis

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Abstract

Neurocysticercosis (NCC) is caused by the larval form of the pork tapeworm Taenia solium and is the commonest parasitic infestation of the central nervous system. NCC is usually managed medically but in some instances surgery is required. Magnetic resonance imaging (MRI) and computed tomography (CT) are often able to provide the diagnosis of NCC in most patients with intraventricular and parenchymal cysts but in patients with hydrocephalus without any cysts, the diagnosis is confirmed by the presence of cysticercal antibodies in the serum. Surgery is usually recommended for intraventricular cysts, hydrocephalus, large cisternal cysts, large parenchymal cysts and when the diagnosis is not certain on imaging studies. For intraventricular cysts, endoscopic surgery is the procedure of choice as it is minimally invasive. For incompletely excised cysts and cysts or granulomas in locations such as the spinal cord, medical treatment with steroids and albendazole is recommended. Hydrocephalus is treated with a ventriculo-peritoneal shunt but shunts in these patients suffer from frequent obstructions and require multiple revisions. The outcome for patients with intraventricular and parenchymal cysts is usually good but for those with hydrocephalus associated with cisternal or racemose cysts and with cysticercotic meningitis, the mortality is high.

Keywords: Cysticercosis, Endoscopy, Hydrocephalus, Outcome

1. Introduction

The larval form (cysticercus) of the cestode Taenia solium or the pork tapeworm causes cysticercosis. Man is an accidental intermediate host who becomes infected by consuming eggs in the gravid proglottids of the adult tapeworm shed in the stools of a carrier of the worm. The natural life cycle has the pig as the intermediate host, man being the definitive host. Cysticercosis is endemic in several under-developed regions of the world except the predominantly Muslim countries of Saharan Africa and Middle East. Larvae of T. solium lodge selectively in certain organs of the body such as the subcutaneous tissues, eyes, muscles and the brain. When they infect the brain it leads to the condition referred to as Neurocysticercosis (NCC).

NCC is the commonest parasitic infestation of the brain and it is estimated that nearly 50 million people worldwide suffer from this disease. Approximately, 50,000 individuals with NCC die each year due to the disease. Seizures or epilepsy is the commonest manifestation of NCC affecting nearly 80% of patients with NCC. A recent study from south India based on a survey of over 50,000 people reported that NCC was the cause of a nearly a third of active epilepsy in the community. Calculations on the basis of this figure would suggest that, in India, there are one million patients with active epilepsy due to NCC.

The mainstay of therapy of NCC is medical and symptomatic. Cysticidal drugs such as praziquantel and albendazole are also used in the treatment of NCC. Surgery is reserved for selected patients with NCC and plays a minor role in the management of the disease.

2. Pathology of NCC

Cysticercus cysts can reside in different intracranial and intraspinal compartments. Most often they reach the parenchyma of the brain through the blood stream. Once they reach the brain, the larva undergoes degeneration at varying rates ranging from a few weeks to several years. The larva lodged in the parenchyma goes through four stages namely, (i) vesicular; (ii) colloidal; (iii) granular-nodular; and (iv) calcific stages. Usually the vesicular or live stage does not cause symptoms. As the process of degeneration begins, the host mounts an inflammatory response to the cyst and this leads to symptoms such as seizures. The other three degenerating stages of the parenchymal cyst can all cause seizures and the colloidal stage can occasionally produce symptoms and signs of a mass lesion producing raised intracranial pressure (ICP) with or without focal neurological symptoms and signs. If the number of cysts in the brain is numerous, then their degeneration can lead to severe edema and severely raised ICP causing headache, vomiting, altered sensorium and even death. This form of NCC is termed...
“cysticercotic encephalitis”. Although most patients suffering from NCC in Latin America and other endemic countries harbour several cysts in their brain, nearly two-thirds of the patients in India have only one degenerating cyst – the solitary cysticercus granuloma (SCG) (Fig. 1). SCG is the commonest cause of focal seizures in Indian patients. It is the cause of focal seizures (with or without secondary generalization) in nearly 50% of patients presenting to some Indian hospitals.

Besides the parenchyma, the cysts can also enter the ventricular system where they may cause hydrocephalus by blocking the circulation of cerebrospinal fluid (CSF). Finally, the cysts can reside in the subarachnoid spaces at the base of the brain or in the sylvian fissures. In this location, the cysts lose their scoles and creep along the subarachnoid spaces. This form of the cyst is termed a “race-mose cysticercus cyst”. The symptoms associated with subarachnoid cysts are due to the severe inflammation of the vessels in the subarachnoid spaces which cause infarcts of the brain stem and the basal ganglia.

NCC rarely affects the spinal cord. Intramedullary cysticercus granulomas are occasionally known to cause paraparesis.

3. Indications for surgery

A group of international experts in cysticercosis published a consensus statement regarding the management of NCC. They considered surgical management for NCC in the following situations:

1. Extraparenchymal NCC
   Intraventricular cysts
   Hydrocephalus due to racemose cysts
   Hydrocephalus due to ependymitis caused by NCC
2. Spinal cysticercosis
   Intramedullary
   Extremadullary

In addition to these conditions, the following situations may need a neurosurgical intervention in patients with NCC.

4. Surgery for intraventricular cysts

Patients with intraventricular cysts usually present with persistent or intermittent features of raised ICP. Computed tomography (CT) or magnetic resonance (MR) scan will usually show hydrocephalus as the cause of the raised ICP. Since the cyst within the ventricle has the same density or intensity as the surrounding CSF, it is often difficult to identify the cyst or cysts. Occasionally, the scolex can be seen as a hyperintense dot within the cyst this being diagnostic of a cysticercal cyst (Fig. 2). A cysticercal cyst in the anterior third ventricle can mimic a colloid cyst. Other differential diagnosis includes choroid plexus or ependymal cysts. Special MR sequences such as 3D CISS (constructive interference steady state) or FLAIR may reveal the cysts. However, even with the special MR sequences, the cyst/s might not be visualised and in cases where an intraventricular cyst is being strongly suspected, a CT ventriculogram will clearly outline the cyst or cysts as filling defects in the contrast filled ventricles.

The cysts can be present in the lateral, third or fourth ventricles, the last location being the commonest. These cysts are free floating and can move about the ventricles depending on the position of the head. As the cyst can migrate from one ventricle to another it is advisable to perform an MR or CT just before the planned surgery in order to avoid an inappropriate procedure or approach. A contrast study should be done to determine the presence or absence of associated inflammation. The inflamed cyst wall will enhance and in such patients it might be prudent to proceed with a shunt and treat the cyst with medical therapy. Cysts with inflamed walls are known to be densely adherent to the surrounding ependyma and choroid plexus and attempts at excision might lead to neurological morbidity and a poor outcome.
Fig. 3. A CT ventriculogram in a patient with suspected lateral ventricular cysticercus cyst. Contrast was injected into the right frontal horn. Note the filling defect in the right lateral ventricle caused by the large cysticercal cyst. Through a right frontal burr hole, contrast was injected into the right frontal horn. Note the filling defect in the right lateral ventricle caused by the large cysticercal cyst. The contrast was then negotiated between the tonsils to the foramen of Magendie where the cyst can be visualised. If there are dense adhesions occluding the foramen of Magendie then the procedure might have to be abandoned.

Occasionally, the cysts are densely adherent to the surrounding ependyma and may not deliver easily. In this case, the procedure might have to be terminated and a shunt would be needed to treat the hydrocephalus. Such cysts can be treated with cysticidal drugs such as praziquantel and albendazole. The excision of lateral and third ventricular cysts can be combined with other procedures such as septostomy to create a communication between both the lateral ventricles and a third ventriculostomy to relieve hydrocephalus due to an obstruction in the region of the aqueduct.

One of the frequently raised concerns is the consequence of the rupture of a ventricular cysticercal cyst while excising it. Madrazo et al. considered the rupture of the cysts to be associated with serious consequences and devised a pipette to perform an atraumatic removal of the cysts. However, most surgeons experienced with the excision of intraventricular or cisternal cysticercus cysts have not reported any untoward incidents associated with intraoperative rupture of cysts. Unlike a parenchymal hydatid cyst, rupture of an intraventricular cysticercus cyst does not lead to either dissemination of the disease or an anaphylactic reaction. Usually patients with a single intraventricular cyst do not need any further therapy after excision. However, those suspected to have more cysts should be treated with oral albendazole (15 mg/kg body weight in two daily divided doses) for 2 weeks. A course of steroids (prednisolone or dexamethasone) can be given for the first 5–7 days of albendazole therapy.

The hydrocephalus usually resolves following excision of the intraventricular cysts and shunts can be avoided in over 70% of cases. However, in some cases, the ependymitis resulting from degeneration of the cysts either within the ventricles or in the subarachnoid spaces might lead to persistence of the hydrocephalus necessitating the placement of a shunt.

Although medical therapy of intraventricular cysts has been proposed, the effect of the therapy takes several days to weeks and during this period some patients might deteriorate acutely. Sudden death with intra-third ventricular cysts has been reported.

5. Surgery for hydrocephalus

Hydrocephalus associated with NCC occurs in the presence of intraventricular cysts, racemose cysts in the basal subarachnoid cistern and due to obstruction at the outlet of the fourth ventricle due to basal cysticercotic meningitis and fibrosis or ependymitis even in the absence of cysts. As mentioned above, hydrocephalus associated with intraventricular cysts usually does not require shunt surgery after excision of the cyst or cysts. A ventriculo-peritoneal (VP) shunt is recommended for hydrocephalus associated with other forms of NCC. As for any inflammatory process causing hydrocephalus, NCC associated hydrocephalus is associated with the need for repeated shunt revisions. This complication may be related to high protein or cell content in the CSF and even due to obstruction of the shunt by small cysts or inflammatory exudates. Shunts have also been known to be the cause of migration of fourth ventricular cysts into the third or lateral ventricles due to a siphon effect. Sotelo et al. have devised a new shunt system which reduced the frequency of shunt revisions from 45% to 30%. However, this system does not provide an ideal solution to the problem of frequent shunt obstruction as in some cases it resulted in the conversion of symptoms of raised ICP to those of normal pressure hydrocephalus. As of now, a standard shunt system is suitable for use in patients with hydrocephalus due to NCC. Long-term steroid therapy with daily oral prednisone at a dose of 50 mg
three times a week for up to 2 years has been shown to reduce the rate of shunt dysfunction from 60% to 13%. However, prophylaxis for tuberculosis with isoniazid is recommended as prolonged steroid therapy can predispose a patient to tuberculous infection.

6. Spinal NCC

Intradural and extramedullary cysticercal cysts in the spinal cord are rare causes of spinal cord compression, the former being more commonly reported. Surgery is usually indicated as the diagnosis is uncertain. However, if there is evidence of cysticercal infection from a positive enzyme linked immunotransfer blot (EITB) for cysticercal antibodies in the serum, then surgery can be avoided. Once a diagnosis of intramedullary NCC is made, the patient might be prescribed steroid therapy alone for a maximum of two weeks or albendazole with steroid therapy for two weeks.

7. Surgery for large parenchymal and racemose cysts

Occasionally patients might present acutely with large parenchymal cysts (usually in the colloidal stage of evolution) causing focal neurological deficits and raised ICP (Fig. 4). In such situations, surgery might be required on an emergent basis to reverse the deficits and reduce the raised ICP. Often, the diagnosis of a cysticercal cyst is also not secured without a biopsy as the imaging features mimic those of an abscess or a metastatic lesion. Craniotomy and excision of the cyst often lead to a good outcome.

Similarly, large cysts in the basal subarachnoid spaces such as the suprasellar cistern or cerebellopontine cistern can present with pressure effects on the adjacent neurological structures. In several instances the diagnosis of racemose cysticercosis is not evident on routine MR imaging. Diffusion weighted imaging (DWI) is necessary to distinguish these cysts from the more common epidermoid cysts. These lesions can be excised through an appropriately placed craniotomy. The cysts are often easily delivered but in some cases parts of the cysts densely adherent to neurovascular structures in the vicinity might have to be left in situ.

8. Surgery for atypical SCG

Surgery is nearly never needed for typical SCG presenting with seizures. However, in some patients SCG can present with atypical symptoms such as severe headache or have atypical radiological features. In 7% of patients with SCG, the granuloma might enlarge (referred to as "enlarging SCG") and a differential diagnosis of other pathologies such as tuberculoma or an abscess might necessitate a histological diagnosis. Finally, SCG in locations such as the brain stem might require histological verification. Stereotactic techniques are often required to excise or biopsy these small lesions located in eloquent regions of the brain. However, if there is a high index of suspicion, then serum should be tested for cysticercal antibodies and if found positive, then either symptomatic therapy or treatment with albendazole will avoid a surgical exploration.

9. Surgery for intractable epilepsy associated with NCC

Occasionally, surgery is required to control intractable epilepsy associated with NCC. The cause of the epilepsy is either an epileptogenic scar around a cortical cysticercal lesion such as a granuloma or mesial temporal sclerosis (MTS) associated with NCC. It has been postulated that NCC could cause MTS by various mechanisms such as kindling, seizures due to NCC causing MTS and due to inflammation spreading from an adjacent degenerating cysticercal cyst. In patients with intractable epilepsy associated with NCC the work up and subsequent surgery is similar to that performed for intractable epilepsy in general.

10. Medical therapy

Medical therapy for NCC is not the focus of this article and hence it will be dealt with briefly. In most patients symptomatic therapy alone is indicated. This usually consists of analgesics for headache, anti-epileptic drugs (AEDs) for seizures and steroids to treat raised ICP.

AEDs may be stopped soon after the resolution of a SCG with a low risk of seizure recurrence. However, in patients with calcific residues of the granuloma or multiple granulomas, AEDs have to be continued for several years due to a high risk of recurrent seizures. Steroids are usually used for brief periods of one or two weeks. Prednisolone (1 mg/kg body weight) or dexamethasone (8–16 mg/day in divided doses) is generally recommended. Steroids are used in patients who present with a flurry of seizures and in those with cysticercotic encephalitis. Steroids are also used in the first week of a cysticidal drug regime.

Cysticidal drugs (praziquantel and albendazole) are known to destroy live larval cysts whether in the parenchyma, ventricles or the subarachnoid spaces and possibly hasten the resolution of granulomas. However, therapy with cysticidal drugs is controversial and there is no consensus on the indications for their use. Similarly, there is no agreement or evidence for the ideal duration of therapy with these drugs. Albendazole is preferred over praziquantel for its lower cost and side effects. A dose of 15 mg/kg body weight in 2 divided doses for 15 days is the most commonly used regime.

There are some types of NCC for which cysticidal drug therapy is contraindicated. Patients with cysticercotic encephalitis may have a fatal outcome if treated with cysticidal drugs due to the increase in edema around the degenerating cysts. Cysticidal drugs by causing acute degeneration of the cysts can lead to an exacerbation of the already elevated ICP causing the death of the patient. Such patients are ideally managed only with steroids and anti-edema measures such as mannitol and glycerol.

Fig. 4. A large right temporal ring enhancing mass on the CT of a patient presenting with features of raised intracranial pressure. The mass was excised and the histology revealed a cysticercal cyst with inflammation. This is a colloidal cyst.
11. Complications and outcome

Complications of surgery for NCC are similar to those for any intracranial pathology. However, endoscopic surgery is generally safe. The major complications for endoscopic surgery include intraventricular hemorrhage, fornical damage, CSF leak, seizures and meningitis. These complications occur in less than 5% of patients.

The outcome following surgery for NCC depends on the form of the disease for which the surgery was done. For intraventricular cysts, large parenchymal cysts, large racemose cysts causing mass effect and for biopsy or excision of atypical SCC, the outcome is generally excellent with over 95% of patients making an uneventful recovery. Gouldwell et al. found that 75% of patients with NCC who underwent surgery had improved at a follow up of 3 years. However, shunt surgery for hydrocephalus caused by cysticercotic meningitis is complicated by frequent shunt revisions for blockages or infection in as many as 68% of patients and carries a mortality of 50% on long-term follow up. Mortality is particularly high in patients with basal racemose cysticercosis.

12. Prevention of cysticercosis

Cysticercosis is a disease associated with poverty and poor hygiene, lack of sanitation and an unregulated pork industry. It is an eradicable disease as it has a well-characterized life cycle in which humans are the only definitive hosts. The best prevention strategy would involve the universal use of latrines by the population. Since that is unlikely to be achieved in a short period of time in several developing countries, other short-term measures have been tried and found to reduce the disease burden. The possible strategies to control cysticercosis include health education emphasizing the need for hygienic practices such as hand washing after defecation and before eating, mass chemotherapy for taeniasis and vaccination of pigs.

13. Conclusions

Surgery is infrequently required in the management of patients with NCC. The commonest indication is for the excision of intraventricular cysts. Endoscopic surgery is the procedure of choice for excision of intraventricular cysticercal cysts as it is minimally invasive. Shunt surgery for hydrocephalus associated with cysticercotic meningitis is the next common indication for surgery. Whereas patients with intraventricular cysts have a good prognosis, those with hydrocephalus have a poor outcome with the need for multiple shunt revisions and a high morbidity and mortality. Miscellaneous indications for surgery in patients with NCC include large parenchymal cysts, spinal cord cysts, atypical SCC and surgery for intractable epilepsy. Eradication of the disease is possible but socio-economic factors prevent achievement of this goal in the near future in several under-developed regions of the world.

Conflict of interest

The author has declared no conflict of interest.

References