Managing cerebral and cranial hydatid disease

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DOI: 10.4103/0028-3886.51276

Human hydatid disease, though preventable, continues to be endemic in major sheep-raising countries worldwide, including India and China. Cases of hydatid have been reported from all parts of India and China, but the disease is endemic in Andhra Pradesh and Tamil Nadu, provinces in southern India,^[1] and western parts of China including Sichuan from where Wang et al.,^[2] report their experience of surgical treatment in cerebral hydatid disease in this issue of the journal.^[2] The tapeworm whose larval stage causes hydatid disease in humans belongs to the phylum Platyhelminthes, family Taeniidae, and class Cestoda. Echinococcus is the genus and there are four main species, E. granulosus, E. multilocularis (alveolaris), E. vogelli, and E. oligarthus. E. granulosus is the most common infestation worldwide including India, whereas E. multilocularis is prevalent in the northern hemisphere including China, and the cases reported by Wang *et al.*^[2] belong to this species. The other two species are prevalent in central and south America. Review of recent literature emphasizes global nature of the disease and possible threat of its spread to countries currently free, mostly related to international travel.^[3]

For E. granulosus, man is an incidental intermediate host and domestic dog is the definitive host. The definitive hosts for E. multilocularis are fox and dog and for the other types it varies in different countries and these are mostly wild animals. Ingested eggs hatch in the duodenum and escaping oncospheres penetrate the intestinal wall and enter the mesenteric venules, and liver represents the first filter through portal circulation. Most of the larvae get filtered in liver and form cysts but some escape to the lungs. After the pulmonary filter few reach systemic circulation and lodge in any part of the body including brain, skull, and bones. In a large series of all human hydatid cysts, bone and cerebral hydatid cysts accounted for 1.0-2.6% and 0.5-3%, respectively. In the calvarium the two common locations are the frontal and occipital bones.^[4,5] Common sites in the cerebrum are the hemispheres, cerebellum, and rarely the brainstem, basal cisterns, and ventricles. Parasite grows into cysts in the brain and the cysts are composed of three layers, two formed by the parasite and one by the host. The outer layer, or pericyst, is composed of inflamed fibrous tissue which is very thin in the brain unlike in other organs; the exocyst is an acellular laminated membrane; and the innermost layer, or endocyst, is a syncytium, which forms the germinal layer of the parasite and gives rise to brood capsules, within which larval scolices develop. An intact cyst is filled with clear fluid. Daughter cysts may develop directly from the endocyst, resulting in multicystic structures. E. granulosus cysts are usually single, rounded, follow endogenous reproduction, and produce symptoms by compression of adjacent tissue. On the contrary, E. multilocularis cysts are honeycomblike, have exogenous budding, and tend to invade the host tissue. Debris within the fluid of the cyst, consisting of hooklets and scolices referred to as hydatid sand, has characteristic radiographic and sonographic features.

The cysts can be single (primary cysts) or multiple (secondary cysts) in the brain.^[6,7] The latter were thought to arise from the multiple scolices released from left side of the heart, following cyst rupture in the heart.

Patients become symptomatic as the cysts grow in size and the presenting features may include raised pressure, focal deficits, or both. Rarely seizures can be the presenting feature. The cysts can rarely get calcified and even infected.^[8-12] In general, unilocular cysts are caused by E. granulosus and the multilocular cysts by E. multilocularis. Multilocular cysts have a more progressive and malignant course and a poor prognosis. Only 42% of operated patients for multilocular cysts achieve good long-term palliation.^[13]

The diagnosis of hydatid cysts is based upon clinical suspicion, particularly in the endemic areas. Positive Casoni's test is diagnostic but a negative test does not rule out the diagnosis. The test can be negative in solitary cerebral hydatid cyst. Serological tests, enzyme linked immunosorbent assay, or indirect hemagglutination, all being more sensitive can be positive in 85% of patients.[14,15] Radiological findings depend upon the type of infestation. Solitary cysts with scolices are very diagnostic.^[16] E. multilocularis lesions may show a variety of findings reflecting the pathology. These cysts in E. multilocularis tend to be small, multiple with areas of solid masses, and may also contain areas of calcification. Rarely the lesion may mimic a primary brain tumor.^[17] Cranial intradiploic hydatid cysts show a honeycomb-like appearance with expansion of the calvarium.

Surgical treatment remains the preferred management for active hydatid disease and the aim of the surgery is to excise the cysts in toto without rupture since this can cause spillage of live scolices and anaphylaxis, a serious complication. Other complication of spillage of live scolices is late recurrence of the disease. Cranial hydatid with many daughter cysts, is difficult to excise without spillage and recurrence is an inevitable complication.^[18,19] It is true of vertebral hydatid disease also. It may be advisable to administer a short course of albendazole before surgery to reduce the risk associated with spillage of cyst contents during surgery. As for surgery on cerebral lesions, one is advised to turn a large scalp flap overlying the lesion based on imaging, followed by careful handling of the cyst with meticulous technique and follow the Dowling-Orlando technique. Adventitial layer around the cyst in the brain is usually thin and it can be separated around the cyst and a saline irrigation around the cyst allows the cyst to extrude itself out. In majority of cases this method of hydrostatic pressure extraction ensures cyst removal without rupture. Thin capsule, especially toward the ventricle and adhesions, may make it difficult for intact removal in some cases. Multiple cysts especially of the E. multilocularis type and the location of the cyst in the brainstem make it difficult to remove them without rupture. In such cases chemical agents can be injected into the cyst to kill viable scolices and 3% saline is useful.

In brainstem regions cyst can be emptied first and then removed since Dowling method is not possible in such a location. Entire operative field should be irrigated with hypertonic saline, which is scolicidal. Wang team used 10% hypertonic saline successfully without recurrence in the short follow-up period of their cases. Cysts located in deeper areas can be removed piece meal and may be followed by prolonged chemotherapy. In deep-seated lesions Wang team removed the lesions piece meal, without recurrence since these lesions of alveolar type may not contain fertile scolices. Wang team successfully excised four cysts en mass and two piece meal with good neurological recovery. Liver disease was responsible for two deaths later. In a recent study, injection of 0.4% chlorhexidine into the cysts (hepatic) for five minutes had been shown to kill all the live scolices and there was no recurrence over a follow-up period of two years.^[20] However, this method is yet to be tried in cerebral and cranial hydatid. One has to study the deleterious effects of 0.4% chlorhexidine to the brain, if that were to get spilled over accidentally during the operation. Effective scolicidal agent is useful especially while removing the cysts containing fertile scolices.

Medical treatment may be the choice if patients are not eligible for surgery. Albendazole results in disappearance of up to 48% of cysts and a substantial reduction in size of the cysts in another 28%.^[14,15] Praziquantel increases serum concentrations of albendazole fourfold and can be used in combination. The duration of antihelminthic treatment could be up to five months or longer. Antihelminthic therapy is beneficial and indicated in patients with spillage or in those patients with disease in other parts, like liver.

Hydatid disease causes considerable mortality and morbidity and a large population around the world is affected by the disease. Surgical removal of the clinically symptomatic cysts and treatment with antihelminthics are only palliative. The best strategy of any public health policy should be prevention and control of the disease. Each country needs to find the species of hydatid causing the disease and the definitive and intermediate hosts responsible in the life cycle of the parasite.^[21] In some countries both forms of the disease, granulosus and alveolaris, may be prevalent. As for India the definitive host is the domestic dog and the intermediate hosts are sheep, goat, and cattle. In a study on the incidence of hydatid infestation in intermediate hosts in Kurnool the positivity of goats, sheep, and cattle was 16.96, 21.75, and 60.94%, respectively. The positivity in dogs in Kurnool region was 33.3%. These facts emphasize the need to control the disease in intermediate hosts and definitive hosts to contain the hydatid disease. Similar steps are needed to control cysticercosis also. Killing of stray dogs humanely and reducing the dog population, surveillance of dogs and periodic test of the stools, and treating them if found infected and preventing from access to raw offal at slaughter houses and farms will be effective to control the menace of hydatidosis. Many countries such as New Zealand, Australia, and China have been successful in this regard and India is yet to catch up.^[22]

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Accepted on 30-03-2009

Source of Support: Nil, Conflict of Interest: None declared.