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Article

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Brain hydatidosis: review of the literature

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Abstract: Hydatidosis is a common disease worldwide. The causal agent may compromise any Organ of the body, the cerebral location is infrequent. The infection is caused by the larval form of Cestodo Echinococcus granulosus. The man is an accidental intermediary host for food consumption or water contaminated with eggs present in animal feces. We present a review of the literature. At the imaging level, the disease has a classic characteristic consisting of single, usually unilocular and less frequent multilocular, intra-axial and more frequently hemispheric cerebral lesions, compromising the vascular territory of the middle cerebral artery by the hematogenous dissemination of the parasite.

Key words: Echinococcosis, Brain neoplasms, Cysts, Neurosurgery

Introduction

Hydatidosis (echinococcosis) is a global zoonotic parasitic disease produced by the larval phase of tenia Echinococcus (1, 2, 3). There are two types of Echinococcus infections: Echinococcus granulosus is the most common type, whereas Echinococcus multilocularis is less common but more invasive, generally with malignant behavior (1, 2, 3, 4, 5, 6, 7, 8).

Most cases of cystic echinococcosis are found in endemic areas such as Australia, New Zealand, the Mediterranean, the Middle East, and South America, and cases of alveolar echinococcosis are found in North America, Central Europe, Russia, China, and Turkey (6).

The overall incidence of organ infestation is highest in the liver reaching 50% to 77%, followed by lungs with 8.5% to 43%, and 10% found in other organs such as brain, muscles, pericardium, kidneys, ocular orbit, tibia, and bone marrow (4, 6, 7, 8).

What is neurohidatidosis?

Central nervous system (CNS) involvement is rare and occurs in approximately 2- to 3% of all cases, and accounting for only 2% of all lesions occupying the intracranial space (1, 3, 4).

Neurohidatidosis is more frequent in children and young adults (approximately 50 to this affects more men than women (62%) from 21- to 40 years -old in the general population (3, 4, 8).

The cyst affects the CNS in 1% of cases and is usually diagnosed in childhood. It has been reported a prevalence of 60-93% in children with a male predominance (5, 8).

Cycle

The life cycle of the parasite requires two hosts: a definitive host and an intermediate host. The main definitive hosts are mainly dogs for E. granulosus and foxes, jackals and coyotes for E. multilocularis, which generally maintain the wild cycle. The main intermediate hosts are sheep for E. granulosus and wild rodents for E. multilocularis (6, 7).

The adult worm lives in the intestine of the definitive host, which expels eggs in feces (infective form) that are ingested by the intermediate host. After the ingestion of eggs, the enzymes in the intestinal tract digest the outer lining of the eggs and the embryo is released. The embryos have small hooks and pacifiers with which they attach to the mucosa of the intestine, perforate the wall and disseminate through the circulatory and lymphatic system (1, 4, 5, 7, 8), and then to the liver, lungs, brain and other tissues. The embryo develops into a cyst within which protoscolices are produced. The cycle is completed when the final host ingests the larval form (5, 6, 7, 8).

Humans may be infected incidentally as intermediary hosts after ingesting the eggs of the parasite directly by contact with a final host or indirectly with contaminated food or water (6, 7).

Histopathology

Histopathologically the E. granulosus cyst wall consists of a germinal inner layer (endochisis) and a laminated outer layer (exocyst). The host reacts to the cyst by forming a capsule of connective tissue (pericyst), vascularized that provides nutrients for the parasite. In the brain, the pericystic layer is thin because and exhibits minimal inflammatory reaction. E. granulosus cysts are usually solitary and commonly supratentorial, located in the distribution of the terminal branches of the middle cerebral artery, usually temporo-parieto-occipital (1, 8)

E. granulosus cysts are usually unique and spherical, well demarcated from the surrounding tissue (which does not present major inflammatory changes), and have smooth and thin walls; they can reach a large size as a result of slow but continuous growth (7, 8). The cystic lesions are homogeneous

with liquid content like that of cerebrospinal fluid, glycolipoproteins, carbohydrates, amino acids and metacestode metabolism salts. Some of its components, such as albumin and immunoglobulins are derived from the host. In the CNS, cysts can grow mainly from the direct implantation of oncospheres or from the metastatic dissemination of a visceral cyst (1, 6, 8).

In contrast to the cyst of E. granulosus, the cyst of E. multilocularis grows by external budding of the germinal membrane with progressive infiltration of the surrounding tissue. Cysts are small, tend to aggregate in clusters resembling a bunch of grapes and proliferate rapidly eroding brain tissues; they have little fluid within a semi-solid structure and many small vesicles embedded in a calcified stroma. They are commonly surrounded with connective tissue edema, and cause a severe immune reaction in the host. They usually do local and distant metastases, especially in the liver, lung, marrow, and brain. The extent of injury to brain tissue is characterized as "crab claws", which is extremely dangerous for the human body (4, 9, 10).

Multiple brain hydatid cysts are rare. Most of the cases are due to surgical, spontaneous or traumatic rupture of a viable primary lesion with implantation of the scythe within the cerebral parenchyma. These lesions are called secondary and are infertile as they lack breeding and scolex capsules. It is believed that multiple cysts can also grow after rupture of a cyst located in a left heart cavity or large arterial vessels (4, 8, 9). In the literature, there are 81 patients reported with multiple cerebral hydatid cysts: 77 by Al Zain et al., 5 by Tu'zu`n et al., and 2 by Buktea et al. (3) Multiple primary cyst without any clinical or radiological evidence of hydatidosis in other parts of the body are rare and are usually result from arterial embolism secondary to the ingestion of multiple larvae (6, 7, 8). In both species, the larva of the parasite can be found within the cysts (7, 8, 9).

Spinal hydatidosis is rarely seen and occurs in less than 1% of all cases of hydatid disease. Thoracic disease is more common with mainly vertebral and paravertebral involvement (1).

Clinical manifestations

The clinical manifestations of neurohydatidosis are closely related to their location. The lesions may remain asymptomatic until they become quite large (10).

The main symptoms are headache, vomiting, nausea, and other nonspecific symptoms such as hemiparesis, seizures, altered visual field and gait disorders; all of them varying with the intracranial location of the lesions. Neurological signs may also manifest as result of increased intracranial pressure (8, 11).

The most common clinical manifestations of spinal hydatidosis include paraparesis (62%), paraplegia (26%), back pain and radicular pain (55%), hypoesthesia (36%), and loss of sphincter control (3%) (8, 9, 10, 11).

Diagnosis

The diagnosis of the disease hydatidosis is based on serological tests and imaging techniques. Immunological data are irrelevant for diagnosis, and the most reliable methods

are neuroradiological imaging and histopathological examination (6, 7, 8, 10, 12).

Image Techniques

Diagnostic methods useful for human hydatidosis include ultrasonography (US), computed tomography (CT) and magnetic resonance imaging (MRI) (13, 14, 15, 16). (Figure 1).

CT (as well as MRI) is essential to the diagnosis of cerebral hydatid cyst. CT sensitivity ranges from 90% to 97%. MRI adequately shows the main characteristics of the hydatid cyst (6, 8, 12, 13).

Although neurohydatidosis can locate in any part of the brain, there are most often found in both hemispheres, particularly in the territory of the middle cerebral artery. In younger patients, skull X-rays may demonstrate suture separation, unilateral and widening of the vault, or erosion of posterior clinoid processes (1, 6, 7, 8, 9, 10).

The evaluations of hydatid cysts by cranial CT and MRI reveal a solitary parenchyma with homogeneous, most commonly supratentorial, spherical and large cysts with well-defined borders, and peripheral edema. The density of cyst fluid is the same as that of cerebrospinal fluid (8, 12).

Although the lesion may cause extrinsic compression of the ventricle system with posterior hydrocephalus, there is associated edema as seen typically in abscesses and cystic tumors. The lesion does not improve after the intravenous administration of contrast material, and calcification is extremely rare (6, 7, 8).

On non-contrast CT, the cyst wall is isodense or hyperdense to the brain tissue (7, 8).

In MRI the cyst wall usually shows a low intensity border in both T1 and T2-weighted images. In T2-weighted MRI, the presence of a hypointense border has been described as a characteristic of hydatidosis (6, 8).

MRI is the preferred imaging modality when spinal hydatidosis is suspected. The walls of the cyst are thin and regular, with no septa. The presence of a markedly hypointense cyst wall in T1-weighted and T2-weighted images, and the absence of enhancement of the wall with gadolinium, are characteristic of hydatid disease (6, 8, 13, 14).

It is believed that MRI should be considered as the imaging aid of choice in planning for surgical approach because of the ability to obtain coronal and sagittal images; and also, because it reveals the anatomical relationships of the lesions to the grooves and ventricles (7, 8, 10, 15).

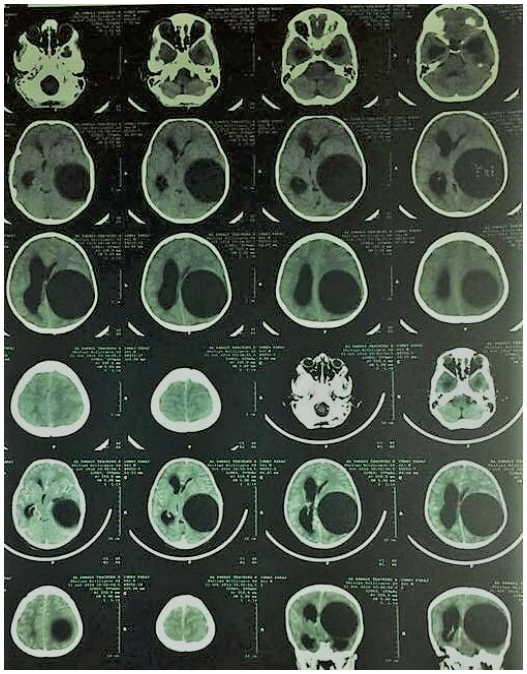

Figure 1 - Cerebral CT scan showing cysts of hydatidosis

In imaging tests, cerebral alveolar hydatidosis are shown as solid, calcified lesions with surrounding edema, in the form of cauliflower. This pattern has also been reported in other studies and may help distinguish cerebral alveolar hydatidosis from other brain masses, particularly in cysticercosis, coenuresis, toxoplasmosis, fungal infections, brain abscesses and oligodendrogliomas (10, 12, 13, 16).

Experimentally it has been shown that a cyst takes from 3 to 16 months to grow to 1 cm in diameter (12).

Serological analysis is in most cases negative. The advantage of histopathological examination is that reveals the degree of infestation of the parenchyma around the cyst that contributes to the enlargement of the process. Although radiological characteristics are characteristic, the definitive diagnosis of hydatidosis is based on the histopathological examination of the surgical specimen (6, 11, 12, 13, 15).

Serology

The test of choice for the serological diagnosis of hydatidosis is based on the detection of IgG antibodies against cyst fluid derived from native or recombinant B antigen subunits, either by enzyme-linked immunosorbent assay (ELISA) or in immunoblots. Antibodies are found up to 92.2% against a new 32 kDa EPC1 antigen of the germ layer of the hydatid cyst of E. granulosus, compared to 84.5% against the B antigen (11, 12, 13, 14, 15, 16).

E. granulosus has been shown to induce both cellular and humoral immune response, therefore increasing the number of CD8 in peripheral blood (10, 14, 15, 17).

In other laboratory tests, the globular sedimentation rate is always high and peripheral eosinophilia could not be a significant feature. Lumbar puncture I usually avoided since changes in cerebrospinal fluid are not specific and the risk of complications due to increased intracranial pressure is high (14, 15, 18).

Evidence for the fixation of the complement (Weinberg) and the reaction of Casoni (immediate hypersensitivity skin test) is limited; Weinberg is positive in 40 to 70% of patients, and false positives are frequently obtained with the Casoni reaction. (14, 15)

Measurement of specific immunoglobulin E circulation is not a diagnostic test but may be used to monitor the response to medical treatment (14, 15, 17)

The release of histamine from basophils and particularly the degranulation tests used to detect IgE binding to basophils, are more sensitive than common tests and may be used to confirm diagnosis in endemic regions (13, 14, 15).

Differential diagnosis

The differential diagnosis of cerebral cystic echinococcosis includes brain abscess, cystic tumor, arachnoid cyst, and encephalic cyst. Hydatid lesions can be differentiated from brain abscess and cystic tumor by the absence of edema around the lesion and mural nodules. Other cystic lesions such as arachnoid cysts and brain cysts are not spherical and are not surrounded by brain matter (7, 8, 15, 16).

The differential diagnosis of cerebral alveolar echinococcosis includes gliomas, metastases, tuberculomas and fungal infections. The diagnosis should be suggested by evidence of a primary hepatic foci, clinical history, patient clinical manifestations, geographic location with high prevalence of infection in the host, and laboratory data (include information about CSF, Western blot, or PCR before surgery (10, 11, 14, 15)

Lack of transparency or sclerosis within adjacent bone in the absence of disc disease in vertebral hydatid disease may help to differentiate from tuberculous spondylitis. (1)

Treatment

The treatment of this infection consists of surgical intervention, medical treatment, and chemotherapy (19, 20, 21, 22).

Medical treatment with mebendazole, albendazole and praziquantel have shown good results in the hydatid cyst of liver and abdomen. Few data have been published in the medical treatment of intracranial hydatidosis with respect to drug penetration through the blood-brain barrier and the intracranial hydatid cyst membrane, however reports of isolated cases showed complete disappearance of multiple intracranial hydatid cysts with albendazole therapy at a daily dose of 10 mg/kg three times daily for 4 months (7, 14, 15, 20, 21, 22).

Nonetheless, surgical removal of the cerebral hydatid cyst remains the treatment of choice. The rate of intact removal of an intracranial hydatid cyst is about 60 to 70%. Intraoperative rupture of the cyst produces a high risk of recurrence and can be fatal due to the dissemination of scolex, which can lead to replantation, anaphylaxis, and chemical meningitis (14, 15, 21, 22, 23).

Medical treatment with albendazole is beneficial in cases of recurrence, rupture of an intraoperative cyst, giant or multiple hydatid cysts, or both before and after surgery. Long-term follow-up of patients with intracranial hydatidosis in whom rupture of the cyst occurred intraoperatively confirm the excellent prognosis reported previously (8, 12). Currently the Dowling-Orlando technique is the technique most used in the surgical treatment of hydatid cysts. When the Dowling technique is combined with microsurgery, large cerebral hydatid cyst can be successfully removed without rupture (14, 15, 21, 22, 24).

The correct treatment of spinal hydatidosis and neurological compromise is laminectomy with removal of the cyst to achieve local decompression. However, cysts that compromise the spine may not be safely cleared, therefore the risk of recurrence is up to 40%, even after extensive surgery (4, 21, 22, 23, 24).

Currently, surgery is the accepted method for the treatment of cases of cerebral alveolar hydatidosis, but chemotherapy is mandatory after the surgical intervention (10) (Figure 2).

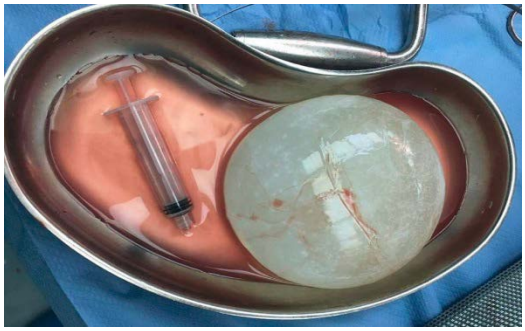

Figure 2 - Appearance of a typical cyst at removal

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