

Case Report

Giant Cerebral Hydatid Cyst in Children

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Abstract

Hydatid cyst is a parasitic disease caused by infection with *Taenia echinococcus* that is endemic to many areas of the world, but due to the current ease of worldwide travel may be seen also in non-endemic areas. The disease commonly affects the liver and the lungs, but may also involve the central nervous system (CNS).

INTRODUCTION

Hydatid is a Greek word meaning “watery cyst” [1]. Hydatid disease is a parasitic and zoonotic infection caused by a tapeworm belonging to the genus *Echinococcus* and family *Taeniidae* at the mature stage (cestodes) or larval stage (metacestodes) [2]. The genus *Echinococcus* consists of four main species: *E. granulosus*, *E. multilocularis (alveolaris)*, *E. vogeli*, and *E. oligarthrus*. The most commonly reported species causing human infection is *E. granulosus*.

The life cycle of *E. granulosus* involves two hosts. Dogs, wolves, and some other carnivores are definitive hosts; adult worms 3-mm to 6-mm length live in their proximal intestine without causing serious health problems, making these primary hosts vectors for later transmission. The eggs of the worms are released from the body of these hosts via feces. Intermediate hosts are generally sheep and cattle that ingest the eggs, where they ultimately enter their gastrointestinal tracts. Parasitic infections by these larvae could lead to cyst formation in different organs, resulting in complications and even death. In most cases, sheep and cattle that are infected are slaughtered before symptoms appear; the infected organs then are given to other primary host animals, continuing the life cycle. Humans can become infected through contact with dogs or other primary host animals; drinking water and eating vegetables can also be contaminated with the eggs of one of these four *Echinococcus* species.

Neurological (brain) involvement occurs in about 1% to 3% of hydatid cysts [3,4], which is rare. Cerebral hydatid cyst accounts for space-occupying lesions of the brain in about 1-5% of such lesions in endemic areas and in 0.2-2% of this cases total population [5]. Central nervous system involvement is associated with a variety of clinical signs and symptoms and significant morbidity and mortality.

The complications of hydatid cyst can be reduced by timely diagnosis and treatment. This paper presents two pediatric cases of giant hydatid cyst.

Case 1

A 12-year-old boy was admitted to our center, with complaints of weakness on the right side of his body. Increased tendon reflexes papilledema, right hemiparesis (2/5), and an extensor plantar reflex in right side were observed during the examination. MRI revealed a brain cyst with a diameter of 8.5 cm and without edema in the left hemisphere (Figure 1). According to clinical symptoms, a craniotomy was performed in which the cyst was ruptured immediately after opening the dura mater and during incision of the cortex.

The cyst fluid was rapidly aspirated and the cyst wall was removed (Figure 2). The patient's hospital stay ended without complications; and albendazole was prescribed at discharge. Six months after surgery and despite continuing oral administration of albendazole (10 mg/kg/day), the patient was referred due to impaired balance and ataxia. On MR imaging, multiple cysts were detected in the brain stem (Figure 3). Regarding the infected site and the clinical symptoms of the patient, the possible postoperative complications and recurrence were explained to the patient's parents, with a recommendation of reoperation and aspiration of the cyst. Albendazole administration was continued



Figure 1 MRI image in the first cases shows cystic mass with homogeneous liquid similar to CSF.



Figure 2 Postoperative ruptured cyst wall.



Figure 3 MRI image in the first case six months after surgery shows the formation of four cysts in the posterior fossa.

after the parents were unwilling to consent to further surgery. Unfortunately, the patient died one year later from aspiration pneumonia and pulmonary complications.

Case 2

A 10-year-old boy was referred to our center and hospitalized with a headache of approximately 50 days' duration and vomiting for fourteen days, followed by a tonic-clonic seizure. During examination, tendon reflexes were intensified on the right side and the plantar reflex was extensor.

On MR imaging, a brain cyst with an immense pressure effect and shift was observed in the left hemisphere of the brain; the intensity of the cyst's fluid was similar to that of the CSF in both T1- and T2-weighted images (Figure 4 A and B). Surgery was performed due to the severity of the clinical symptoms, and the cyst was completely removed without rupture after craniotomy, opening the dura mater, incision of the cortex, and releasing of the adhesions (Figure 5). No postoperative complications occurred, and the patient was returned to his normal activity despite having a large subdural effusion on CT scans one month later (Figure 6).

DISCUSSION

The hydatid cyst caused by *E. granulosus* is a major endemic health problem in the regions bordering the Mediterranean Sea, North Africa, South America, the Baltic regions, Spain, Greece,

Turkey, Portugal, the Middle East, Australia, New Zealand, and the Philippines [1]. The adult worm lives and produces eggs in the intestines of definitive primary hosts often, carnivores such as dogs and foxes. These eggs are released in the intestine of the host and excreted within the feces to the environment. The eggs immediately become infective and resistant to environmental factors and remaining viable for a long time in different environments. Humans can be infected by eating food or drinking water containing these eggs. When the eggs are transmitted into the human digestive tract, digestive enzymes lead to the opening of their chitin shell, releasing the larvae. These larvae adhere to the intestinal wall, penetrate the sub-mucosal jejunum, and enter into the veins or lymphatic system, thereby spreading through the lymphatic and port circulation. Through the veins and intestinal lymph nodes, the larvae first enter the liver, which effectively filters about 75% of them. Some of these larvae pass through the filtering of liver and reach the right ventricle of the heart through the inferior vena cava (IVC) and lungs. Potentially 15% of the larvae are trapped in the lungs. The larvae that pass through the liver and lungs can enter the systemic blood circulation and then circulate to any location in the body; some potential locations include the peritoneum, spleen, kidney, heart, brain, spine, skeleton, and muscles. The involvement of other organs may be rare [6], but no area of the body is protected from infection once the larvae pass through the filtering of the liver and the lungs [3].

Another reported vector for transmission in the inhalation of



Figure 4a MRI T1.

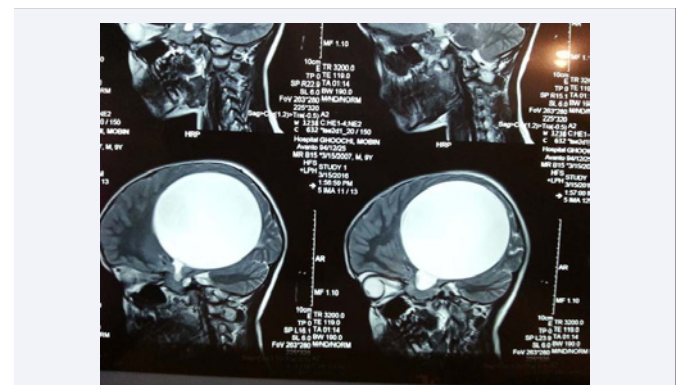


Figure 4b and T2 weighted program of the second case shows a very large cyst ; its fluid features are quite similar to CSF.



Figure 5 The cyst during surgical removal from the brain, and at the end of surgery.

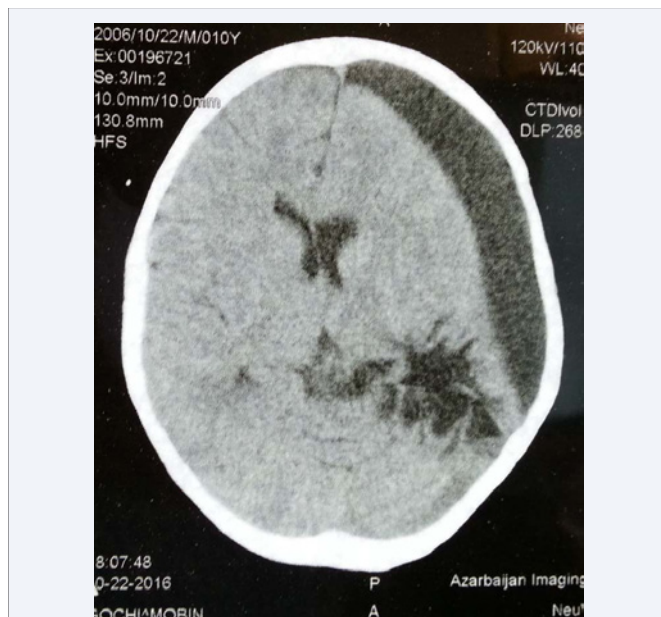


Figure 6 CT-Scan one month after surgery shows subdural effusion without residual cyst.

dust contaminated with parasite eggs; in this case, the larval shell may be opened in the lungs and enter into the bloodstream [4,7].

When the larvae are established in the intermediate host tissues, cyst formation begins. It should be noted that many of them (over 90%) will be eliminated by the host immune system [1]. The cyst comprises three layers: the innermost layer that creates the germinal epithelium (fertile layer), daughter cysts, and scolexes. If the scolexes enters the digestive system of the definitive (primary) host, adult worms could be created there. In addition, if these cysts are ruptured spontaneously or during surgery, the scolexes could be transported to other areas of the body, causing secondary cysts. Over time, the number of these daughter cysts and scolexes are increased inside the cyst, leading to cyst growth. The intermediate cuticular layer is the middle layer of the cyst and consists of amorphous and laminated chitin that is produced by the inner layer of the parasites. The outermost part of the cyst is the adventitial layer, which is synthesized by the host (unlike the two inner layers). This layer is created as a result of the immune response of the cyst-embedded tissue, which consists of fibrous tissue and contains a large number of

eosinophils and inflammatory cells. Therefore, a greater number of tissue reactions to the cyst lead to an increased thickness of the adventitial layer. The thickness of this layer is often high in the liver, low in muscles, and moderate in the brain tissue; it is often nonexistent in bones [8]. In areas where the thickness of this layer is small or does not exist, spontaneous or surgical cyst rupture becomes more likely.

The cysts formation rate in various tissues is different depending on the tissue resistance, and the cyst can grow 2-3 cm in diameter per year [9,10] and this rate can be even quicker in children [11]. Cyst growth rates are higher in compressible tissues such as brain and lung. The cyst growth rate in the brain is three times higher than in the liver [12]. In the brain, it takes 5-16 months to form a 1-cm cyst [9] that then grows 1-10 cm per year [13].

Hydatid cysts affect the brain in 1-3 % of cases [3,4], and any part of the brain could be involved [9]. The higher frequency of involvement in the brain hemispheres is due to the spread of the parasite through the bloodstream. These cysts are often single [14] and commonly occur in the watershed area of the middle cerebral artery (MCA) due to their embolic nature [4], especially in the parietal lobe [15]. Multiple primary cysts transmitted by blood are rare; rather, multiple cysts are usually secondary brain cysts caused by spontaneous, traumatic or procedure-associated rupture of primary cysts. The primary cyst may also rupture in an area such as the left ventricle, leading to diffuse spread to different areas of the body (such as the brain) and the formation of multiple cysts [14].

70-80% of CNS hydatid cysts are found in children [8,9], particularly in boys [9]. The primary reason for the higher prevalence of hydatid cysts in the cerebral hemispheres in children is related to patent ductus arteriosus in heart [17], allowing direct right-to-left shunting of blood flow in the heart and evading filtering by the lungs.

A patient with a hydatid cyst may not have any significant symptoms for a long time due to the slow, gradual growth of the lesion. As the cyst increases in size, signs of increased intracranial pressure (ICP) will appear, including headache, nausea, vomiting, blurred vision, and double vision. Depending on the cyst formation site and the effects of resulting pressure, other symptoms could be seen, including hemiparesis, gait disorder, seizures, and cranial nerve paralysis. Finally, large cysts can cause cerebral shifts and herniation, leading to reduced levels of consciousness and, over time, death if not addressed.

A plain radiograph of the skull does not usually lead to definitive diagnosis, but may show the localized skull bulges and symptoms of increased ICP (such as destruction of the posterior clinoid process) or pathologic calcification. CT scanning and MR imaging are techniques suitable for the diagnosis of cerebral hydatid cyst [5]. MR imaging is preferred because of multi planar imaging of the lesion, showing better anatomical details and lack of complications related to radiation [3]. Both methods show perfectly round cystic lesions in the cerebral hydatid cyst (*E. granulosus*); the cyst manifests on imaging with similar features to those of CSF, without surrounding edema. Contrast agents do not enhance the cyst's features on imaging.

In patients with suspected lesions related cerebral hydatid cyst, other organs of the body (especially the liver and lungs) should be investigated for the presence of cysts.

Despite reporting rare cases of cerebral hydatid cyst treated by pharmacological therapy with albendazole [18,19] long-term studies have demonstrated that cerebral hydatid cysts should be surgically removed [17]. The purpose of surgery is to remove complete cysts without rupture, as ruptured cysts (similar to our first case) can lead to secondary cyst formation through releasing of scolexes, potentially resulting in serious complications and death.

If the patient's clinical condition is stable, it is better to administer albendazole for a short period before surgery. This reduces the number of viable scolexes in the cyst, with accompanying reduction in the risk of accidental rupture during surgery. Cyst removal should also be easier with reduction of pressure inside the cyst.

Surgery must be performed without touching the cyst; Dowling-Orlando's method is widely used for this purpose [20]. During operation, a large craniotomy is needed at the proper site to prevent excessive pressure or pull (which could cause a rupture). Since most cysts are close to the cerebral cortex or exactly under the dura mater, the dural opening procedure must be carried out carefully. After opening the dura mater and cortex incision (if necessary), the cyst is adequately exposed (about $\frac{3}{4}$ of the diameter of the cyst), and a soft catheter (usually Foley) is carefully inserted below the cyst after releasing brain-tissue adhesions and cyst wall. Then, the cyst is gradually removed by slowly injecting the area around it with water to create hydrostatic pressure. In addition, using some maneuvers, such as increased ICP provided by the anesthesia care team, placing the head in a dependent position, and using gravity and pressure from the hand(s) on the brain tissue adjacent to cysts, could help to remove cyst. In patients with multiple cysts, the first objective is surgical removal of the largest cyst [20]; other cysts are treated in the same session or in subsequent sessions.

The major complication of hydatid cyst surgery is intra operative accidental rupture of cyst. In this case, the cyst fluid should be quickly aspirated, and the surgical field should be rinsed by hypertonic saline, silver nitrate, or 1% formalin [21].

In some cases, the cysts are established in critical or deep areas of the brain and surgical removal is not feasible. In other scenarios, anesthesia and surgery is impossible due to the patient's medical condition. For these situations, the cyst could be aspirated [22] or a Gamma Knife surgery can be used [23].

Drug treatment should also be administered in this group; patients who cannot undergo surgery due to the position of the cyst or medical problems, who have multiple cysts in various organs or tissues, or who have an intraoperative ruptured cyst. For this purpose, albendazole or mebendazole can be used, which inhibit glucose uptake in the larvae [17,24]. Praziquantel combined with albendazole could also be used to increase serum levels of albendazole by a factor of four [25]. The minimum recommended duration of drug treatment is 6 months [26]. In the case of inoperable brain lesions, this time has been recommended to be longer and even until the end of life in some cases [27].

Therapeutic prognosis depends on the number, size(s), and site(s) of the cyst or cysts as well as the outcomes of surgery. The most important factor in prognosis is intact removal of the cyst at surgery [28]. In patients with multiple lesions or with lesions in critical areas, surgical complications will be higher, because cortical incision(s) are often required in critical or several areas of the cerebral cortex.

CONCLUSION

Although hydatid cysts commonly affect the liver and lungs that may rarely involve the brain. Brain involvement by hydatid cysts requires surgery that is associated with serious complications and sometimes even death. Since the disease could be transmitted to humans from dogs through orofecal route, administration of anti-worm medication for domestic dogs, avoiding feeding the viscera of slaughtered animals to dogs and other animals and burying them, and paying attention to personal and food hygiene could reduce the rate of Echinococcal infection that leads to the development of these cysts.

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