Giant Cerebral Hydatid Cyst; A Rare Case Report

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Key Clinical Message

Here we describe a 13- year old patient with the presentation of fever and abdominal pain. He had a history of 2 years headache and seizure. In MRI, a primary cerebral hydatid cyst was evident. The diagnosis of hydatid cyst should be considered in children with mentioned characters in endemic regions.

Keywords: Hydatid cyst, Brain, Iran, Surgery

1. INTRODUCTION

Hydatidose, is an important zoonotic disease caused by dog tapeworm *Echinococcus granulosus*. Dogs are definite hosts for the parasite and passes eggs with their faces. Humans are accidentally infected through ingestion of contaminated food or water with dog faces or by direct contact with dogs [1,2]. The disease is common in the Mediterranean countries, the Middle East, Australia and New Zealand [3]. Cysts occur most commonly in the liver or lungs [1]. The intracranial involvement is rare and with incidence 1-2% of all cases of hydatidosis [4]. We describe a case of a giant cerebral hydatid cyst in a 13 years old boy.

1. CASE REPORT

A 13-year-old boy, presented to our hospital with abdominal pain and fever. The pain was constant, non-radiation, worsened with light activity. He has any others gastrointestinal symptoms. The patient had no history of previous hospitalization and did not take any medication. The patient lived in a rural zone.

In admitting time, the patient was febrile (body temperature 40˚c), blood pressure 110/75 mm Hg, heart rate 92 beats/min, breathing rate18 bpm. A general physical examination revealed no abnormality.

1. DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS AND TREATMENT

The complete blood investigation performed with results of: a white blood cell count of 7400 cell/mm3; hemoglobin 12.4 g/dL; platelets 275000 cells/mm3. Laboratory examinations revealed Alkaline Phosphatase levels (548 IU/l) which was high. Also blood and urine culture, serological tests and C– reactive Protein (CRP) were performed. For all of these tests results were negative. Urinalysis and other laboratory work-ups were normal. Erythrocyte sedimentation rate (ESR) was 16 mm/h. Chest radiography, electrocardiogram and ultrasonography (US) of the Abdomen and Pelvic were normal.

Cranial Computed tomography (CT) revealed a left parieto-occipital hypodense, cystic, well-demarcated (Figure 1). Magnetic resonance imaging (MRI) showed the mass to contain membranous structures and with cerebrospinal fluid (CSF) intensity in the left hemisphere, paraventricular area (Figure 2).

Based on the cranial Imaging findings, differential diagnosis of other cystic lesions like abscesses, large granulomas, cystic gliomas, epidermal cysts and arachnoid cysts, arising from the brain were considered. However, imaging and serological findings confirmed the diagnosis of hydatid cyst. A wide parieto-occipital craniotomy was performed. The dura was dissected (Figure 3) and the cyst expelled without rupture (Figure 4). The patient recovered well after the Surgery. Pathological examination of the specimen confirmed the diagnosis of hydatid cyst.
Treatment with albendazole was started when the patient was discharged from the hospital, and he also received Dilantin (phenytoin sodium). The patient fallowed up 1 year after the surgery.

1. DISCUSSION

Echinococcosis is a parasitic disease caused by the larval stage of a tape worm *Echinococcus*. Dogs and other canids are definitive hosts and sheep,goat and other herbivores are intermediate hosts. Humans get infected accidentally through oral-fecal route by consumption of vegetables or water contaminated with parasite eggs or via direct contact with an infected dog. There are four species of this genus can infect humans: *Echinococcus granulosus* (causing cystic echinococcosis), *Echinococcus multilocularis* (causing alveoloar echinococcosis), *Echinococcus oligarthus* and *Echinococcus vogeli* (which cause polycystic hydatid disease). After ingestion of eggs, they hatch in the small intestine sub mucosa and enter veins or lymphatic vessels [5], then disseminate in to organs such as liver, lung and other organs. The most common sites are the liver (75%) and lungs (15%) followed by the spleen, kidney, heart, bones and brain (10%).
Central nervous system hydatidosis is rare, and is usually diagnosed during childhood [6].
In our report, the patient was young and had no disease; living in a rural area and contact with dogs were his only risk factors. Consideration of hydatid cyst in person with fever and history of contact with dogs is necessary in endemic areas.
Brain hydatid cyst also is classified as primary (single) or secondary (multiple), the primary cysts are formed as a result of direct infestation of the brain without involvement of other organs and the secondary multiple cysts results from spontaneous, traumatic or surgical rupture of a solitary cranial cyst [2]. In the present case cysts identified only in the brain, so the brain was a primary focus for hydatid cyst.

Hydatid cyst see in anywhere of the brain, its most commonly located supratentorially, in the middle cerebral artery territory [6], the presented case have a single supratentorially cyst in the left parietal and occipital lobe, and paraventricular area (Figures1-4).

Intracranial hydatid cysts usually present with headache, vomiting, and seizure due to raised intracranial pressure and brain compression. In physical examination, papilledema and neurological deficit may be presented [7]. This case only presented generalized abdominal pain, fever with a history of seizure and headache. In general examination no abnormality was found.

A combination of tools including imaging techniques and serology are necessary for diagnosis of a patient with hydatidosis. For diagnosis of cystic echinococcosis (CE), imaging techniques (CT scan and MRI) are the most reliable methods while serological tests which detect the specific antigens of *E- granulosus* are used for verifying the imaging results [8]. In our patient hydatid serological test and CRP were negative and the other laboratory tests were normal.

CT scan of cranial hydatid cysts showed an intraparenchymal spherical cystic lesion with distinct borders. Also the cyst fluid was isodense with CSF. MRI imaging showed a low signal intensity rim of the cyst wall, while the cyst content had signal intensities similar to CSF [9].

In our case CT scan revealed a left parieto-occipital hypodense, cystic, well- demarcated, round lesion causing shift to the right. MRI imaging showed a mass consists of membranous structures with CSF intensity in the paraventricular area of left hemisphere.

Although in small or inoperable brain cysts, medical therapy has shown promising effects but, surgery remains the golden treatment by which cysts can be removed without rupture and results a complete cure. Chemotherapy with two benzimidazoles (ABZ or MBZ) is indicated for inoperable patients with primary liver/lung echinococcosis and for patients with multiple cysts in two or more organs [10].

Dowling technique is the most effective surgical method for the removal of cerebral hydatid cysts without causing rupture [11]. For Dowling-Orlando technique head puts lower than the operation table. Large craniotomy flap can be made depending on the size and site of the lesion. Surgery area of brain covering cotton soaked with warm normal saline to prevent spillage in case of rupture. Then the cyst removes and dura closing water tight, bone flap puts back and patient dresses after wound closure [2, 12]. After craniotomy and prior to the incision, the surgical field must be cleaned with scolicidal solution to prevent recurrence because even a minimal spillage can lead to new cysts formation (1ml of cyst fluid contains 4000000 scolices) [13, 17].

Albendazole has been used successfully for treatment of hydatid cyst in brain [14], for prevention of secondary hydatid disease. Use of benzimidazoles (ABZ or MBZ) before surgery can reduce the risk of recurrence of CE. Chemotherapy is more effective among younger patients. For treatment of CE, oral dosage of 10-15mg/kg/day of Albendazole in a course of 1 – month course separated by 14-day interval can be prescribed. Three courses are routinely suggested, and more than six usually will be unnecessary. The usual oral dosage of Mebendazole is 40-50mg/kg/day for at least 3-6 months [15].

In summary, intracranial hydatidosis is rare and more affects pediatric age group. It may misdiagnose as intracranial cyst, so in differential diagnosis of intracranial cyst especially in endemic areas, age of the patients will be helpful. Total surgical remove of the cysts without rupture is still the treatment of choice in cerebral hydatidosis.

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AUTHOR'S CONTRIBUTIONS:

All authors, M Gh, A Sh , F.RY and SA.H are equally contributed to the design, analysis, and presentation and critically revise of this study.

B.G is surgeon of patient. M Gh is specialist in infectious disease and involved in study design.

A Sh: Involved in study design, writing, submission and revision.

DECLARATION

-Ethics approval and consent to participate: applicable

-Consent for publication: Written informed consent was obtained from the parents of patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

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193Figure legend

194Figure 1: pre operation CT scan revealed a large cystic left parietal and occipital lobe,
195paraventricular area.
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197Figure 2. T2-weighted axial MRI of the brain shows a large cystic left parietooccipital lobe,
198paraventricular area.
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201Figure 3. After opening the dura, an intra-cranial cystic mass was determined. The lesion was
202removed gross totally.

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204Figure 4. The cyst removed in toto after operation. The cyst appears with creamy and
205germination of daughter cysts