

## Primary Giant Cerebral Hydatid Cyst in an 8-year-old Girl

### Abstract

Echinococcosis, also called hydatid disease, is a parasitic disease that passes from animals to humans. Literature reports suggest very rare cases of cerebral hydatid cysts. Brain involvement with hydatid disease occurs in 1%–2% of all *Echinococcus* infections. In this report, we aim to emphasize the presentation of such an isolated primary cerebral hydatid cyst, discuss its radiological features, Emergency department management, inpatient medical management, referral to neurosurgery, consequent operative procedures, postoperative care, and outcome.

**Keywords:** *Albendazole, cerebral, Echinococcus, hydatid cysts, surgical excision*

### Introduction

Echinococcoses are parasitic diseases of major public health significance. Human infection results in chronic disease with variable prognosis, serious medical, social and economic consequences for vulnerable populations. According to a recent study, the geographical distribution of *Echinococcus* spp. infections is expanding and becoming an emerging and re-emerging problem in several regions of the world including southern South America, Mediterranean littoral, southern and central parts of the former Soviet Union, central Asia, China, Australia, and some parts of Africa.<sup>[1]</sup> The larvae of *Taenia Echinococcus* cause hydatid cyst. Dogs are the definitive host where mammals such as sheep and cattle are the intermediate hosts.<sup>[2]</sup> Humans usually get the infection through food contaminated by dog feces (containing the eggs), besides the direct contact with dogs.<sup>[3]</sup> Brain involvement with hydatid disease occurs in 1%–2% of all *Echinococcus* infections. Most cases of intracerebral echinococcosis diagnosed the world were in children.<sup>[4]</sup>

### Case Report

We report this case, presented to King Abdullah Specialist Children Hospital a Tertiary Pediatric Hospital part of King Abdulaziz Medical City, Riyadh Saudi Arabia. The consent of the patient enrolled, obtained and the study approved by our institution for publication. An 8-year-old

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Syrian girl presented to our ED with progressive blurred vision over 20 days, headache that was getting worse 1 week before her presentation, sudden bilateral eye exophthalmos more intense on the right side.

There was no history of vomiting, neurological deficits, seizure attacks, or abnormal movements. No changes in behavior or activity, she had no fever or weight loss. Clinical examination on presentation was normal, neurological examination was normal no neurological deficits, ophthalmic examination showed bilateral eye exophthalmia with decreased vision acuity, blurring of optic margins suggesting papilledema. Diagnostic workups performed.

Brain computed tomography (CT) showed a large cystic lesion in the left frontal intra-axial, measured approximately 9 cm × 9 cm showing multiple peripheral solid nodules, midline shift, and brain edema observed with basal cistern effacement. Early uncal herniation noticed. We referred her urgently to neurosurgery team [Figure 1].

Keeping in mind rare presentation of Brain hydatid cyst, chest X-ray done for her and did not show any lesion in lungs. Ultrasound of the abdomen was normal. CT chest and abdomen were done and were normal.

The patient admitted to the Pediatric Intensive Care Unit (PICU) for monitoring,

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she was active, conscious, and stable. After a written consent from the parents/guardians, the patient sent to the operation room and the cyst removed by the Dowling-Orlando technique with the aid of gravity without rupture [Video 1]. Histopathological examination confirmed the hydatid cyst stating a translucent cyst weighing 500 g and measuring 20 cm × 15 cm × 12 cm (*ex vivo* measurement which is expected to differ from the *in vivo* radiological measurement of 9 cm × 9 cm) in our search this is the largest brain hydrated cyst in the literature [Figure 2].

She was successfully extubated on the same day then sent back to PICU; medical therapy was started as Albendazole, Vitally she was stable and clinically with Glasgow coma scale of 15/15, communicating and moving all her limbs normally. Day 1 postoperative, she had one seizure attack, which was brief with visual hallucinations. The seizure managed by diazepam and phenytoin responded well. Brain CT done for the patient on same day showing improvement in the mass effect and the midline shift, but no changes in the brain edema [Figure 3].

All laboratories postoperative were normal including the electrolytes, complete blood count, and coagulation profile. After 3 days in PICU, we transferred her to the ward in stable condition, kept in hospital for 16 days for monitoring, on albendazole as medical treatment, then discharged home with follow-up.

## Discussion

Cerebral hydatid cyst classified as primary or secondary. Primary cysts usually formed because of direct infestation of larvae in the brain without the involvement of other organs. Patent ducts arteriosus and patent foramen ovale have been the proposed pathological factors for isolated cerebral hydatid disease.<sup>[5]</sup> Our patient Echocardiography revealed no abnormality.

Recognition of neurological signs, abnormal raised intracranial pressure, and imaging features by the physician is lifesaving. In Erashin *et al.* they observed 18 out of 19 cases presented with raised intracranial pressure, four of them had seizures.<sup>[4,6]</sup> In our patient, we were able to recognize characterized symptoms and signs of neurological involvement, besides imaging appearance of apparent multiple peripheral solid nodules in the cystic lesion.

Common magnetic resonance imaging findings of hydatid cyst showed smooth, thin walled, spherical, homogeneous cystic lesions.<sup>[7]</sup> The appearance of the cyst fluid was similar to that of cerebral spinal fluid. The CT scan showed cyst wall to be isodense or hyperdense to brain tissue. The overall calcification was rare (<1%).<sup>[8]</sup> In our patient, the cyst showed similar characteristic features; however, the other characteristics of differential diagnosis of cerebral cystic lesions (pyogenic abscess, cystic astrocytoma, pencephalic cyst, and arachnoid cyst) also considered here. The shape of

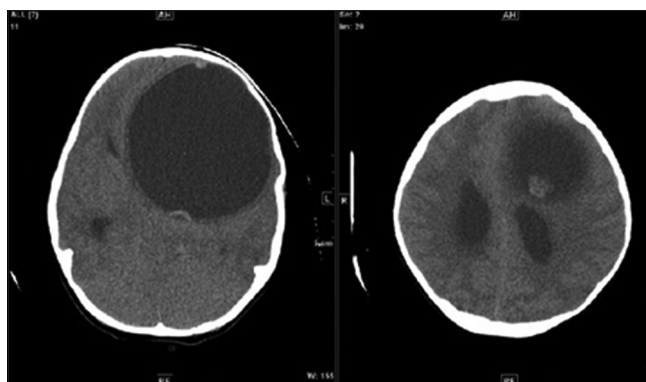


Figure 1: First brain computed tomography showing large left frontal intra-axial cyst

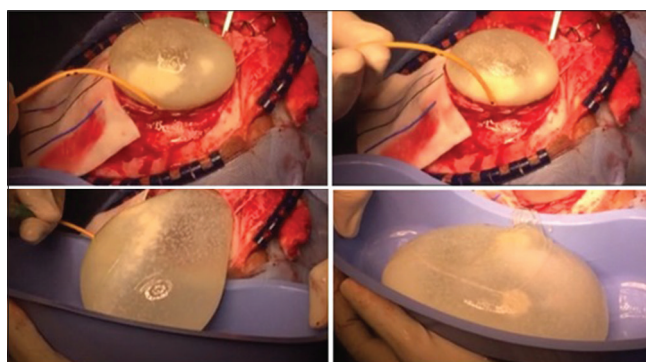


Figure 2: Successful removal of the cyst by Dr. Azzubi

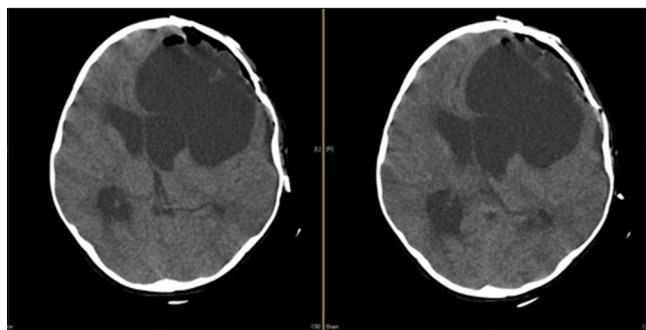


Figure 3: Brain computed tomography day 1 postcraniotomy with surgical changes and interval improvement

the cyst was spherical, so we excluded both pencephalic cyst and arachnoid cyst, which are usually irregular, brain abscess, and cystic astrocytoma excluded by the absence of significant rim enhancement or brightly enhancing mural nodule.<sup>[9]</sup> Although cerebral hydatid cysts are very rare, they considered as one of the differential diagnoses when evaluating a patient with a cystic brain disease.

Surgery is the treatment of choice. With our patient, we used the Dowling-Orlando technique, which is the most preferred technique to avoid the rupture of the cyst. Other techniques had described in literature, such as fluid aspiration, but they appear to be dangerous with a higher chance of the cyst rupture causing complications such as anaphylaxis or parasite seeding risking recurrence.<sup>[10]</sup>

Medical therapy is also important it is effective in sterilizing the cyst, decreasing the risk of anaphylaxis, and reducing the recurrence rate. Albendazole is the drug of choice, but in the absence of Albendazole, Mebendazole can use as an alternative therapy. Albendazole is dosed 10–15 mg/kg/day in two divided doses for around a 1-month period.

The optimal duration of medical treatment of hydatid disease is uncertain and depends mainly on the nature of the cyst being single or multiple, with or without significant leak or rupture, how the cyst had removed, i.e., completely intact or not.<sup>[11]</sup> In our patient, the huge cyst successfully and completely removed without rupture. Thus, a short course of 4-week treatment with albendazole initiated. Eosinophil count was normal for her, and *Echinococcus* serology was negative (<1:16), this can be explained by the lack of interaction between host tissue and the cyst, the cyst was completely intact, and no minimal leak occurred. The patient seen in the neurosurgery clinic after 1 month, vitally stable, thriving well, no complaints, no neurological deficits, and with the normal neurological examination.

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#### Conflicts of interest

There are no conflicts of interest.

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