

## Case Report:

# A Case Report of Primary Brain Hydatid Cyst in a Child



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## ABSTRACT

**Background and Importance:** Hydatid cyst is endemic in India, with different clinical presentations according to the site of involvement in the body. However, its occurrence in the brain without the involvement of other body parts is rare.

**Case Presentation:** An eight-year-old female child presented with right-sided limb weakness and slurring of speech. The diagnosis of primary brain hydatid cyst was made with Magnetic Resonance Imaging (MRI) of the brain. The cyst was removed completely, resulting in improved post-operative slurring of speech and limb weakness.

**Conclusion:** In countries where hydatid disease is endemic, brain hydatidosis should on top of the differential diagnoses for intracerebral cystic lesions and managed as per hydatid protocol until proved otherwise. This approach is critical to prevent rupture and dissemination.

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## Highlights

- The patient had a primary brain hydatid cyst. The patient's echocardiography, abdomen, and chest computed tomography scan were normal.
- In our study, the patient was a female child, but in literature, male predominance was present, i.e. 50%-70%
- In our study, the patient did not have a headache and presented with slurring of speech, right Upper Motor Neuron (UMN) (type facial palsy, and right-side hemiparesis).

## Plain Language Summary

Hydatid cyst disease is caused by dog tapeworm (*Echinococcus granulosus*). It rarely involves brain, which is more commonly seen during childhood due to cardiac septal defects. Brain MRI is the investigation of choice for brain hydatid cyst for its management. Treatment of choice is surgery with care during surgery to avoid its rupture. Albendazole is given to the patients to prevent its recurrence and the development of small microscopic cyst anywhere in the body.

### 1. Introduction

In tropical countries, hydatid disease contributes to a major health burden among zoonotic diseases. Hydatid disease is endemic in Libya, Tunisia, Algeria, Iran, Iraq, Turkey, Morocco, Egypt, and India [1]. But today, some cases are seen in non-endemic regions as a result of migration. The most involved organ by the hydatid cyst is the liver (77%). The less frequently involved organs are the lungs (43%), kidneys (2%), heart (2%), brain (2%), eye (1%), spinal cord (1%), spleen, spermatic cord, and soft tissues [2]. The highest prevalence rate is reported in the Mediterranean region, which is around 8% [2]. Hydatid cyst can have spontaneous, traumatic, or even iatrogenic rupture. Thus, misdiagnosis can lead to anaphylactic reactions and dissemination. To avoid this, the brain surgeon should keep a high level of suspicion in mind for brain hydatidosis, when a patient presents with an intracerebral cystic lesion in endemic countries.

### 2. Case Presentation

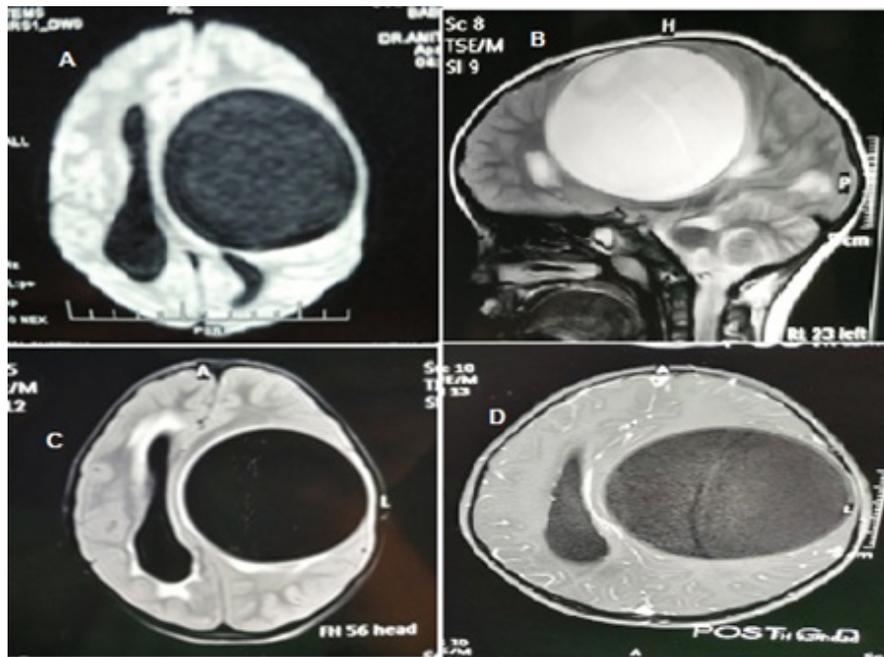
An eight-year-old girl presented progressive right-sided hemiparesis, left deviation of the angle of the mouth, and slurred speech for two and a half months as main complaints. She had no history of headache, seizure episode, or any pet exposure. On examination, there was apparent Left central facial palsy, and power in the right upper and lower limbs were MRC grade 3/5 and 4/5, respectively. A Grade II papilledema (Frisen scale) was detected bilaterally. All other investigations including routine blood test, chest X-rays, chest and abdomen Computed Tomography (CT), and echocardiography were within normal range.

Head CT scan showed a very large intraparenchymal hypodense lesion involving the left fronto-parietal region, with no contrast enhancement and no diffusion restriction on MRI. There was also no associated perilesional edema on brain MRI (Figure 1A-D). After a trial of empirical albendazole, the patient underwent a large craniotomy to remove the cyst. The cyst was removed totally intact via the Dowing's technique (Figures 2, 3A and 3B) [3, 4]. Post-operative contrast-enhanced CT revealed the complete removal of the cyst (Figure 4). After an uneventful recovery, the patient was given a treatment regime of 10 mg/kg/day albendazole for six months and follow-up showed no recurrence during clinical examinations. Chest X-ray and ultrasonography of the abdomen was done at every follow-up session and the brain CT was done at month three post-operation. Complete improvement in the speech was achieved in the first-month post-operation, and complete improvement in right facial nerve palsy and right side limb weakness was achieved in the month three post-operation.

### 3. Discussion

#### 3.1. Pathogenesis

Hydatid cyst is an infectious disease which begins with the entrance of the larval stage of dog tapeworm (*Echinococcus granulosus* and *Echinococcus multilocularis*) in the human body passed on via exposure to a definitive host (generally a dog) or by eating contaminated water or vegetables. Hydatid cyst has two layers, an outer pericyte layer, which is composed of a fibrocollagenous lamellated chitinous layer, and an inner germinal layer, accompanied by brood capsule and hooklets embedded in it. Generally,



**Figure 1.** Pre-operative contrast MRI of the brain showing left-hemispheric hydatid cyst

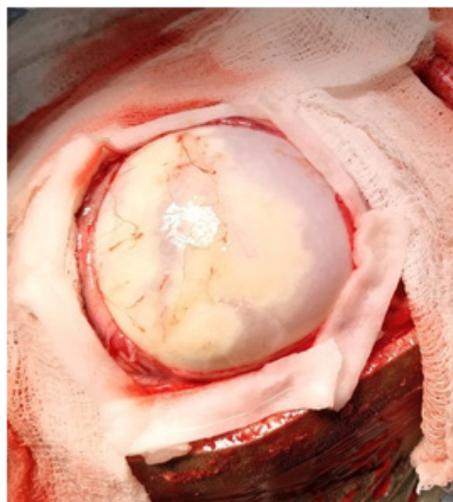
A: T1 axial image; B: T2 sagittal image; C: FLAIR axial image; D: T1 contrast axial image

the cyst remains asymptomatic until an intra-organ mass affects it and it gets ruptured [3].

Brain involvement in hydatid cyst disease may occur as primary or secondary. The former happens when the larva escapes the liver and lung filtration barriers and directly enters into the brain, while the latter develops secondary to a hydatid cyst rupture in another organ with the resultant brain involvement via blood stream.

Primary cysts are most of the times solitary and fertile, but secondary cysts are generally multiple and infertile [4, 5]. The proposed mechanisms for the development of primary brain hydatid cyst are defective immune system within the brain parenchyma, the presence of cardiac anomalies like patent ductus arteriosus, and patent foramen ovale.

The higher affinity of the certain genotype of *E. granulosus* for the brain and variations in class II HLA in normal population leads to a different rate of susceptibility to hydatidosis [1, 5].



**Figure 2.** Intra-operative image of the brain showing the hydatid cyst

The dura is intact.



**Figure 3.** Removal of the intact hydatid cyst

A: The space occupied by the cyst is clearly shown; B: The diameter of the intact cyst 12cm × 10cm × 9cm

The majority of the cerebral hydatid cysts (75%) occur in the pediatric age group, being two to three times more common than in adults [1]. In our study, the patient was a female child, but in the literature, male predominance was found (i.e. 50%-70%) [5]. The most common site in the brain is the parietal lobe [5]. Other uncommon brain regions are the calvarium, eyeball, cavernous sinus, cerebellum, pons, ventricles, and extradural region [6]. Most reported cases had solitary, intraparenchymal cysts in the supratentorial region and in the vicinity of the middle cerebral artery [7].

Particularly in pediatric age, the elasticity of calvarium and high compressibility of brain tissue leads to high growth rate and development of the large size of brain

hydatid cysts [8]. The rate of growth in the brain is usually faster than in any other organ and it is said to be around 4.5 cm/6 months [1]. Tuzun et al. stated that other organs' involvement with brain hydatid reaches up to 80-90% in post-mortem examinations, whereas in clinical practice, concomitant extracranial cysts were rarely demonstrated. The hydatid cysts in other organs are too small to be diagnosed by clinical and radiological evaluations [1].

### 3.2. Clinical presentation

Clinical features vary according to the size of the hydatid cyst and its location. Patients mostly present with headache and vomiting as a result of increased Intracranial Pressure (ICP). Other manifestations are seizures,



**Figure 4.** Post-operative contrast-enhanced CT scan of the brain, showing improvement in the midline shift of the hemispheres due to the removal of the cyst.

**PROTOCOL SCHEME FOR BRAIN HYDATID CYST** <sup>[9,11,15]</sup>

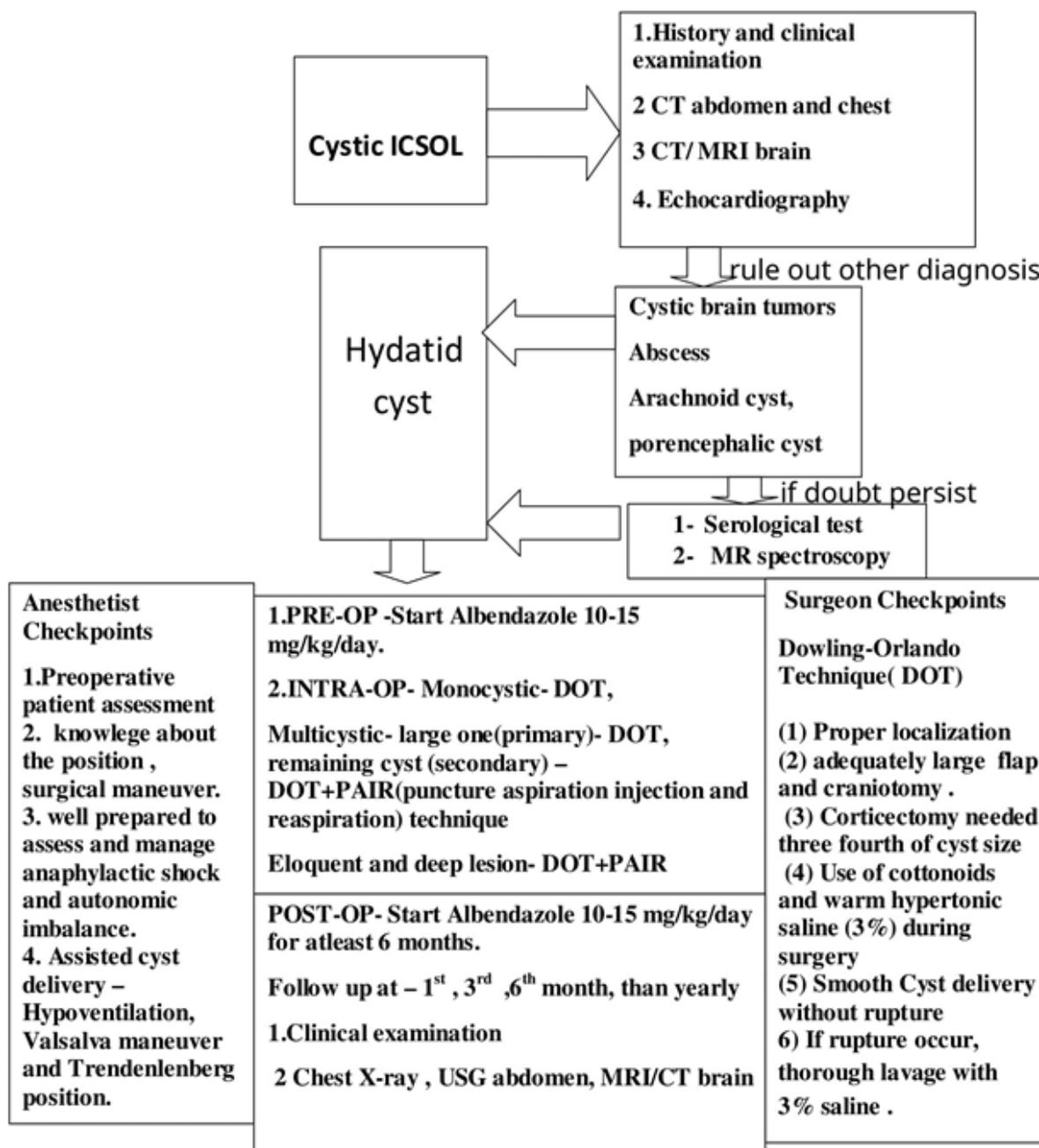


Figure 5. Flowchart of proper protocol scheme to manage brain hydatid cyst



hemiparesis, visual disturbances, speech difficulties, cranial nerve deficit, and ataxic gaits. Papilledema is mostly present at the first hospital visit in children [1]. In our study, the child did not have headache or vomiting and presented with slurring of speech, right UMN type facial palsy, and right-side hemiparesis.

**3.3. Differential diagnosis**

Differential diagnoses of hydatid cyst are the other brain cystic masses in pediatric patients like a pyogenic or fungal abscess, cystic astrocytoma, cystic granuloma, arachnoid cyst, and porencephalic cyst. Pilocytic astrocytoma enhances in CT and MRI and has a characteris-



tic feature which is known as a contrast-enhancing nodule (moral nodule). Although arachnoid cysts occupy an extra-axial location like hydatid cysts, they are not as round as hydatid cysts and are not surrounded by the brain tissue.

Abscesses have a thick wall, which brilliantly enhances with contrast, and the edema surrounding it. In neurocysticercosis, multiple cysts are seen in different stages, and in fact, they have a pattern called "hole-with dot", as the larval scolex could exist as a mural nodule [9].

### 3.3.1. Serology

The cutaneous test described by Casoni, the complement fixation reaction described by Ghedini and Weinberg, indirect hemagglutination, ELISA, and eosinophilia were classically used to diagnose hydatid cyst. Nevertheless, as a minimal response would be elicited within the brain in cerebral hydatidosis, false-negative results may occur frequently [2]. Hence, serologic studies are of little clinical importance in brain hydatid cyst [4, 10].

### 3.3.2. Radiology

Brain CT and MRI are not the only keys to radiological diagnosis, but they are also beneficial for planning the surgery. Brain CT displays hydatid cysts as intra-axial, homogenous cystic lesions with well-defined borders. The cyst fluid is isodense with the Cerebrospinal Fluid (CSF), and the cyst itself is usually round. Non-contrast CT scans display hypodense, well-circumscribed, and circular cysts compared to the brain tissue. The perilesional edema and enhancement of contrast seen with abscesses or cystic tumors are unusual for brain hydatid cysts [11]. MRI displays brain hydatid cysts as hypo-intense on T1 weighted image, and it appears as a hyper-intense region on T2 weighted image with a hypo-intense disc surrounding the cyst. Excellent peripheral enhancement accompanying perilesional edema may be present in active inflammation, although calcifications are only seen in 1% of the cases. Though, alveolar hydatid appear as a contrast-enhancing solid cystic lesion with perilesional edema, and calcification [11]. Measuring reliable markers such as alanine, succinate, acetate and lactate on Magnetic Resonance Spectroscopy (MRS) before and after treatment is an excellent indicator of efficient chemotherapy [2].

## 3.4. Treatment

### 3.4.1. Medical management

Albendazole is currently the drug of choice; it inhibits the uptake of glucose by both the larva and the adult worm. This results in depletion of their glycogen stor-

age which subsequently leads to diminished Adenosine Triphosphate (ATP) production, causing death of the larva and adult worm [5]. Albendazole is effective as it sterilizes the cyst, decreases the anaphylaxis risk, and reduces the rate of recurrence [10]. In some case reports, effective disappearance of multiple brain hydatid cysts with daily doses of albendazole, given three times of 10 mg/kg for a period of four months has been reported [12]. Some studies have also reported the effective disappearance of around 48% of the cysts and prominent reduction in the size of 28% of the cysts following albendazole treatment [2].

It should be noted that mebendazole and albendazole are teratogenic drugs and should not be used for pregnant patients [1]. These drugs are recommended at least for six months [7], and monitoring should be done with clinical and radiological evaluation. In the case of *Echinococcus multilocularis* infection, these drugs should be given to the patients for up to two years. In patients with inoperable lesions, it might be necessary to use the drugs as long as possible, even to the end of life [6]. That way, albendazole is given at the dose of 10-15 mg/kg/day in two divided doses. In case of non-tolerable side effects to albendazole, mebendazole is given 40-50 mg/kg/day [13].

### 3.4.2. Surgical management

Surgery is the treatment of choice for hydatid cyst, and aims to complete excision without rupture, to prevent recurrence and immunological reaction. Many techniques for hydatid cyst excision have been investigated, all of which have reached a consensus on the necessity of an atraumatic removal of the cyst to avoid rupture.

The Dowling's technique has been used as the preferred operative technique for CNS hydatid cysts. During which the delivery of the cyst is done by declining the head of the operating table by 30 degrees and hydro-dissection and Valsalva. This is feasible due fragile adhesions around the cyst wall [14]. Although lowering the ICP by anesthetists is a critical step to provide better access to the cyst, in some cases a small increments in ICP is required to help the cyst being pushed out.

In such cases, hypoventilation is done to increase the PaCO<sub>2</sub> and the Valsalva maneuver may be used in Trendelenburg position [15]. High index of suspicion is required to diagnose an anaphylactic reaction and to manage it by early aggressive surgical treatment and delivering 100% oxygen with epinephrine infusion and massive fluid resuscitation. To prevent and treat the refractory hypotension, prophylactic hydrocortisone supplementation is also rec-

ommended [15]. The proper protocol should be followed as represented in the schematic flowchart (Figure 5).

### 3.4.3. Complications

An intra-operative rupture of hydatid cysts can lead to fatal anaphylactic reaction and a high chance of recurrence [12]. The reported recurrence rate in the brain due to cyst rupture at surgery is reported 40.7% [1]. Mainly reported post-operative complications are hemorrhage, seizures, subdural effusion, porencephalic cyst, hydrocephalus, pneumocephalus, and focal neurological deficits, which sometime may need further intervention [7]. The space left after the excision of large hydatid cysts may lead to fatal complications, such as subdural hematomas, hyperpyrexia, cerebral edema, cortical collapse, or cardiorespiratory failure, in the early post-operative days. In late post-operative days the rate of complication such as subdural effusion and the porencephalic cyst. Cortical collapse associated with mortality is around 0.07 [16]. Majority of cases with neurological deficits improved completely as Hilmani et al. described 90% recovery in the neurological deficits in their case series [5].

## 4. Conclusion

In endemic countries (even in non-endemic countries due to migration), while considering differentials for cystic masses of the brain, the hydatid cyst disease has an important place. Its rupture can be detrimental for the patient, so proper protocol should be followed for complete excision to prevent anaphylactic reaction and dissemination.

### Ethical Considerations

#### Compliance with ethical guidelines

This research was approved by the Ethics Committee of SMS Hospital, Jaipur, India.

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#### Authors' contributions

All authors have contributed equally to the manuscript.

#### Conflict of interest

The authors declare no conflict of interest.

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