Case Report

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Paediatrics Section

Primary Cerebral Hydatidosis in a Child: A Case Report with a Rare Site of Occurrence

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ABSTRACT

Hydatid cyst of brain in an extremely rare entity. A case of primary hydatidosis of brain without any involvement of other areas is even rarer. Most commonly involved areas are liver and lung because of rich vascularity and vicinity to portal circulation. Central Nervous System (CNS) or most importantly cases of hydatid cysts of brain are very rare. This case report is about a 7-year-old girl, from eastern India, who presented with multiple hydatid cysts in cranial cavity. She presented with headache, sudden loss of balance, short time memory loss. On Magnetic Resonanace Imaging (MRI), multiple cystic lesions were noted in cranial cavity. Magnetic Resonance (MR) Spectroscopy findings were suggestive of hydatid disease. These cysts were removed through neurosurgical intervention. The diagnosis was confirmed by gross and microscopic examination. Importance of keeping hydatidosis as a differential diagnosis while working up cases of cystic Space-occupying Lesion (SOL) of the brain in children is evident here.

Keywords: Anthropozoonosis, Brain, Cyst, *Echinococcus*, Hydatid, Neurosurgery

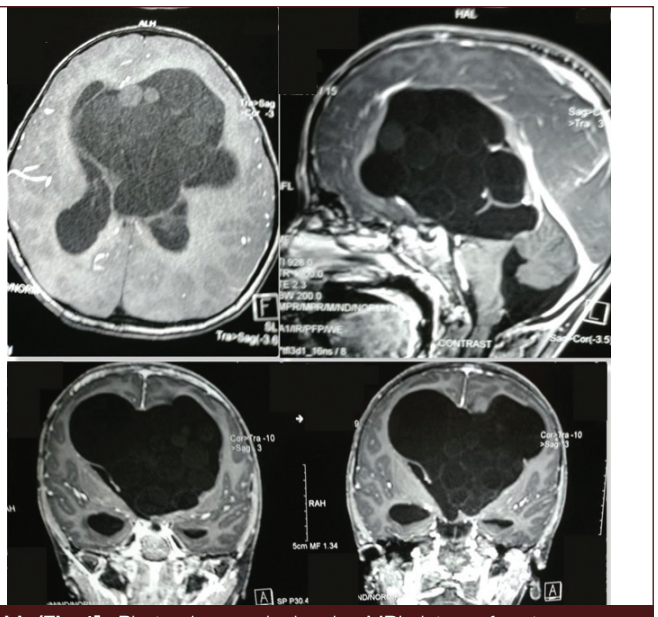
CASE REPORT

A 7-year-old girl complained of having non specific symptoms like headache, vomiting, sudden loss of balance, and short time memory loss for around six months. Conservative treatments by several general practitioners proved futile as the symptoms slowly intensified. On suspicion of a brain lesion, she was referred to this tertiary care centre by a general practitioner.

Her general physical examination was unremarkable, except moderate weight loss. Her routine blood examination revealed no significant abnormality, except for a total count of 15650/cumm. She came from a rural family with poor hygiene practices.

The MRI brain was done suspecting a Central Nervous System (CNS) lesion, revealed hyperintense cystic lesion involving lateral and third ventricles, a marginated small septa was seen in it. Postcontrast study showed no such enhancement; features favouring inflammatory condition likely to be hydatid cyst [Table/Fig-1]. MR Spectroscopy showed reduction in N-Acetylaspartate (NAA) peak with slight increase in choline peaks, presence of lactate and acetate peak was observed. Succinate peak was also observed. While NAA/creatinine ratio was decreased and succinate/acetate ratio was increased. All these features were suggestive of hydatid cyst. MR Angiography was done which revealed mostly normal Middle Cerebral Artery (MCA) and Anterior Cerebral Artery (ACA) with subtle pressure effect caused by the cystic lesion. Rest of the study was unremarkable.

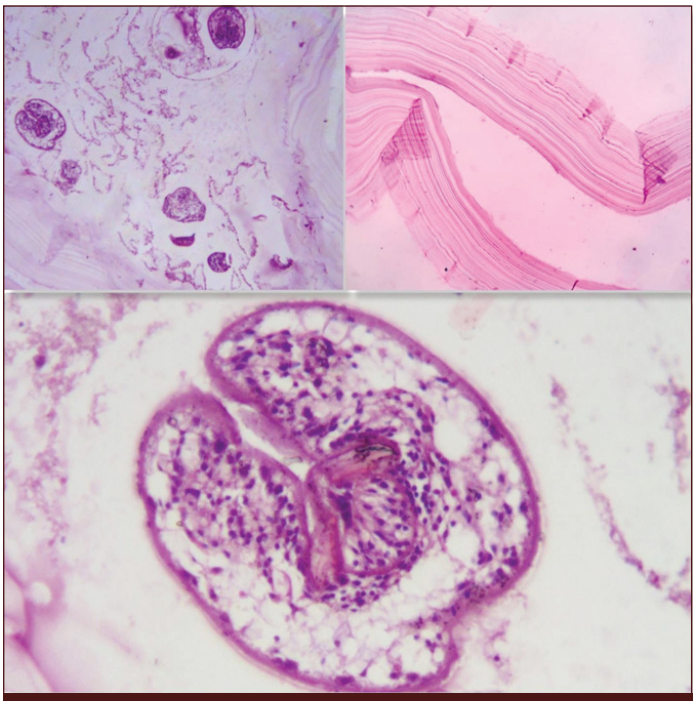
A daily dose of 10 mg/kg Albendazole was prescribed preoperatively for 4 weeks to sterilise the cyst and decrease the cyst wall tension. The surgery was done through Dowling's procedure. After exposure of the brain, the cysts were dissected slowly through hydrostatic pressure and gradual advancement of cotton strips. After successful removal of the cysts [Table/Fig-2], the cavity was irrigated with warm hypertonic saline for its scolicidal effects.



[Table/Fig-1]: Photomicrograph showing MRI picture of cyst.

[Table/Fig-2]: Photomicrograph showing peroperative picture of multiple cyst ball.

The specimen was sent to Department of Pathology for further processing. On microscopic examination, it showed cystic structure with outer fibrous pericyst layer, middle laminated ectocyst layer and inner germinative layer with brood capsule and daughter cyst [Table/Fig-3]. So, it was confirmed histologically as primary cerebral hydatidosis. As there was significant cerebral edema postoperatively, intravenous dexamethasone 4 mg thrice daily was given for three days. As seizures are common both post and preoperatively prophylactic anticonvulsant drug levetiracetam 10 mg/kg was prescribed. Albendazole 10 mg/kg daily was prescribed postoperatively to be continued for 4 months. The next day after operating procedure a Computed Tomography (CT) scan brain was done, which was satisfactory. Her postoperative period was uneventful. The patient was discharged on postoperative day 10.



[Table/Fig-3]: Photomicrograph showing histopathological features of hydatid cyst wall and daughter cyst H&E (100x and 400x).

DISCUSSION

Hydatidosis is an anthropozoonosis, caused by different organism of *Echinococcus* species. Although there are few species involved in the disease like *E. granulosus*, *E. multilocularis*, *E. oligarthrus* and *E. vogeli*. *E. granulosus* is by far the most common species [1]. Only in 1-2% cases it affects CNS [1]. This infection is much more common in children as 50-75% cases involve children [2]. Hydatid cyst is in 60% cases found in liver and in 30% cases in lung [3]. Due to the vasculature of the brain, hydatid cyst is common in supratentorial region than infratentorial region as middle cerebral artery supplies 75-80% of brain [4].

Adult tapeworms live in canine small intestine and, the eggs are found in excreta. The intestine of a canine is the definitive host. It spreads to human, the intermediate host through accidental consumption of food contaminated with dog excreta. In hydatidosis the parasite mainly enters body through ingestion. Interestingly, case of inoculation through bee sting [5] has been documented.

After ingestion the shells of the ova opens due to the acidity of upper digestive tract. Then the hexacanth embryo penetrate mucosa and migrates by the portal system to the liver, where hydatid cysts are formed and may metastasise mostly to lung. In very rare cases it metastasise to brain, periorbital tissue, vertebrae, kidney and pericardium. After inoculation a central cyst develops from the embryo with a thick non nucleated laminated external layer or cuticle. This layer behaves as a semipermeable membrane lined by a thin germinal nucleated membrane rich in glycogen from which brood capsules or daughter cysts arise remaining attached to the germinal layer [4]. Hydatid cysts are filled with fluid rich in scolices which together with the brood vesicles form the hydatid sand [6]. In majority of the cases, the cerebral hydatid cysts are found in paediatric population [7,8]. Most of the cerebral cysts are solitarily and seen in the territory of MCA [9]. Involvement of brain stem is rare [10]. Usually, when small in size, hydatid cyst of brain do not cause major symptoms. But gradually as it increases in size non specific symptoms like headache manifest [11].

It has been observed that cerebral hydatid cysts in paediatric population are usually solitary [8]. Cerebral hydatidosis of paediatric population are primary in majority of the cases [12,13]. Multiple primary hydatid cyst is a rare occurrence and, may form due to rupture of solitary primary cyst or embolisation from cyst of another organ [14,15]. There are two cases of multiple primary cysts from India, as reported by Tanki H et al., [10]. In those cases mean age of presentation was 11.5 years, with a male to female ratio of 5:4. The most common presenting symptom was headache.The [Table/Fig-4] [16-22] shows a compilation of published articles on cerebral hydatid cyst according to location and clinical presentation.

Clinical manifestation of hydatid cyst is variable depending on the location and size of the cyst, as in most of the cases these lesions are clinically silent [23,24]. However, in this report symptoms were sudden loss of balance, sudden loss of consciousness, headache and short time memory loss. High degree of clinical suspicion is key for the timely diagnosis in this part of the world where hydatidosis is endemic.

Author and year	Geographic location	Gender/Age (years)	Multiplicity	Location	Presentation	Treatment
Saqui AE et al., [16] 2017	Morocco	Male, 12	Solitary	Posterior fossa	Headache,vomiting	Surgery+Albendazole
Lakhdar F et al., [17], 2010	Morocco	Male, 37	Multiple	Posterior fossa	Convulsion	Surgery+Albendazole
Bhaskar S et al., [18] 2012	India	Female, 40	Multiple	Posterior fossa	Headache, vomiting	Surgery+Albendazole
Gazzaz M et al., [19] 2000	Morocco	Male, 5	Solitary	Posterior fossa	Torticollis	Surgery+Albendazole
Karakoc ZC et al., [20] 2016	Turkey (Iraqi patient)	Female, 14	Multiple	Bilateral CP angles	Respiratory distress+quadriparesis	Surgery+Albendazole
Fakhouri F et al., [21] 2015	Syria	Female, 5	Solitary	Right cerebellar hemisphere	Convulsion	Surgery+Albendazole
Raynham OW et al., [22] 2009	South Africa	Female, 25	Multiple	Left CP angle	Limbs/face weakness	Surgery+Albendazole
Present study, 2022	India	Female, 7	Multiple	Lateral and third ventricle	Headache, vomiting, sudden loss of balance	Surgery+Albendazole

[Table/Fig-4]: Published literature on cerebral hydatid cyst from various parts of the globe [16-22].
ICP: Intracranial pressure; CP Angle: Cerebellopontine angle

In the majority of the cases serological tests like anti echinococcus antibody (IgG antibody) is positive [25,26]. A round to ovoid well-circumscribed hypodense cystic lesion without pericystic edema is typically seen in CT scan brain [27]. Hydatid cyst appears as hypointense lesion in T1 weighted images and hyperintense with a hypointense halo around in T2 weighted image in MRI, in presence of peripheral inflammatory reaction ring enhancement can also be seen [28,29]. In this particular case, MRI and MR spectroscopy was suggestive of hydatid cyst. However, MR angiography was unremarkable.

Surgical intervention remains the best choice of treatment. The goal of surgery is to remove the cyst without spillage of the live scolaces causing anaphylaxis or recurrence after surgery [30,31]. In this case, there was multiple cysts in the cranial cavity. All the cysts were removed without spillage. In Dowling's technique, hydrostatic pressure is used to dissect the fragile cyst wall from cerebral parenchyma [32]. The plane between the cyst wall and the cerebral parenchyma is created by cotton patties. A soft tipped rubber catheter is then introduced in the plane and for continuous irrigation. Along with saline irrigation and slow progression of cotton patties or strips, gradually the cyst is dissected out intact [33]. Long-term follow-up have shown that hydatid cysts should always be removed surgically and prognosis is usually excellent [32].

Dowling's technique is widely used as surgical treatment of cerebral hydatid cyst [33]. Pre and postoperative Albendazole may be considered to sterilise the cyst, reduce the tension minimising chance of rupture and, anaphylaxis and reduce recurrence rate [34]. Dowling's technique was later developed by Arana-Iniguez and San Julian [35]. Despite being widely used,the technique has its pitfalls concerning surgical instruments, location and methods [36]. Location of the cyst is of great importance. As it is impossible to reach the cyst without exposing some neural structures unless its just beneath the dura. In some cases, cortical incision and cortical retraction in necessary [36]. Cortical incisions and retraction has been in some cases uneventful and in others has produced transient or permanent deficits [36]. However, in this case the follow-up period was uneventful. Other intraoperative complications can be like adhesion around the cyst wall and bleeding during its removal [36]. These events were not there in this case. In present case, the cyst involved lateral and third ventricles which is a dangerous location surgically [36]. Special attention should be paid on the removal of these cysts as the ventricular wall must be preserved. Tear of intraventricular wall and puncture of cyst could cause leakage of cystic fluid in the Cerebrospinal fluid (CSF) causing anaphylactic chemical meningitis [36].

CONCLUSION(S)

Primary CNS hydatid disease is a rare entity. As this condition only manifests with non specific symptoms high index of suspicion is essential for early diagnosis. Especially in endemic regions prevention of this disease condition should be emphasised. Histopathological examination along with MRI findings are considered as confirmatory diagnosis of hydatidosis.

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