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Unusual Location of Hydatid Cysts in Pediatric Patients

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Key Words
Hydatid cyst, unusual location · Computed tomography · Magnetic resonance imaging

Abstract
Objective/Aims: To emphasize the importance of diagnosis and treatment of unusually localized hydatid cysts in pediatric cases. ***Methods:*** Hydatid cyst patients of two departments were listed who had undergone surgery between January 2001 and December 2008. Of the 7 pediatric patients, 3 were chosen as the ones with unusual localization. Cyst removal with Dowling's technique was performed in 2 cases and total removal of the cyst wall was achieved after cyst aspiration in the other patient. ***Results:*** Two patients did not show any signs of recurrence. Some of the cranial multiple cysts of the patient who had undergone her first surgery in another clinic with cyst rupture were successfully removed in our clinic. Six months later, she was admitted with spinal seedings. ***Conclusion:*** Hydatid cyst removal without rupture should be the surgical goal in all cases. Radiological evaluation is of utmost importance for differential diagnosis. When a cystic lesion is found in the central nervous system on radiological evaluation, hydatid disease must be considered in countries where the disease is endemic and surgery is to be planned emergently especially for pediatric cases with increased intracranial pressure. The study focuses on the strategy for the correct diagnosis and the appropriate treatment of unusually localized hydatid cysts.

Introduction

Echinococcus granulosus is a parasite of dogs, wolves, foxes and jackals. Human beings are affected by the excreted eggs found in the feces of the sick animals. After the development of the embryo, various organs are infested through the migration of the intestinal mucosa. The liver and the lung are the most frequently involved organs with ratios of 75 and 15%, respectively [1–3].

Usually, primary hydatid cysts are solitary and may reach huge dimensions in a long time interval. The cysts are reported to grow slowly; the average growth has been observed to be 1–5 cm per year [4, 5]. This growth rate may be even greater in untreated cases, i.e. up to 10 cm [6].

Central nervous system (CNS) echinococcosis is rare and only 2–4% of the cases with hydatid disease have CNS involvement [7–11]. Brain involvement in hydatid disease occurs in 1–2% of all *E. granulosus* infections [6, 12]. Regarding the lesions found in the deeper areas of the brain, the cysts located in the hemispheres of the brain

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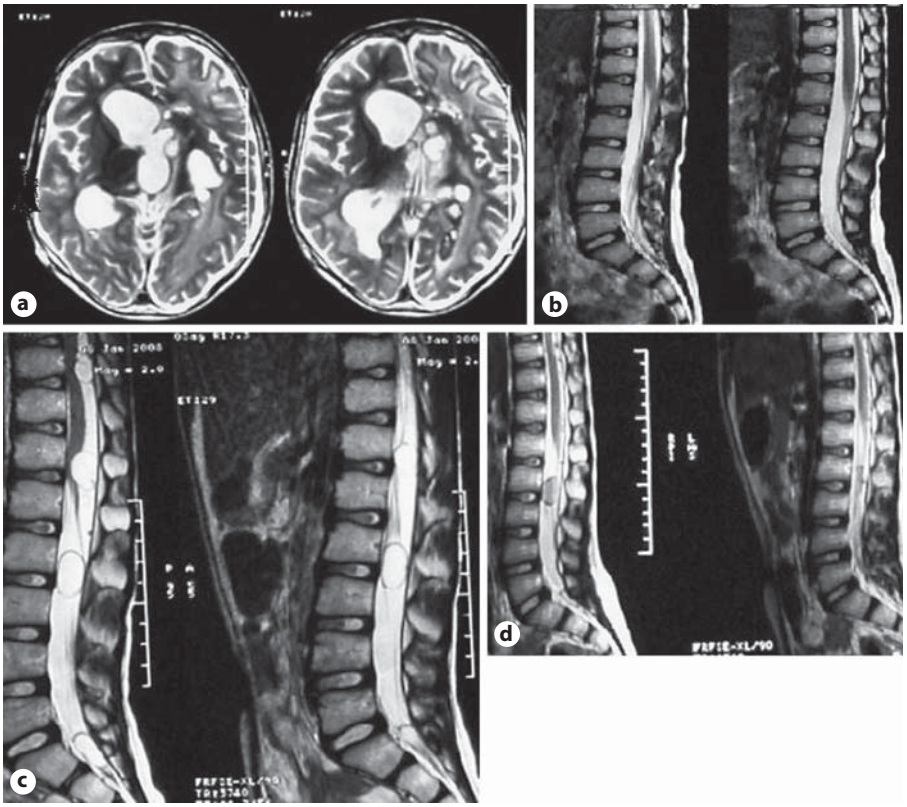


Fig. 1. a Multiple cerebral hydatid cysts. **b** The spinal MRI revealed no pathologic lesions in the spinal column. **c** Multiple spinal seedings of hydatid cysts 6 months after the patient's admittance to our clinic. **d** The last condition after 6 months of albendazole treatment.

are surgically removed considerably more easily. The deeper parts are reported to be extremely rare areas of localization of cysts [13, 14]. Spinal hydatid cyst is rare and its frequency among all hydatid cysts has been reported to be approximately 1% [15, 16].

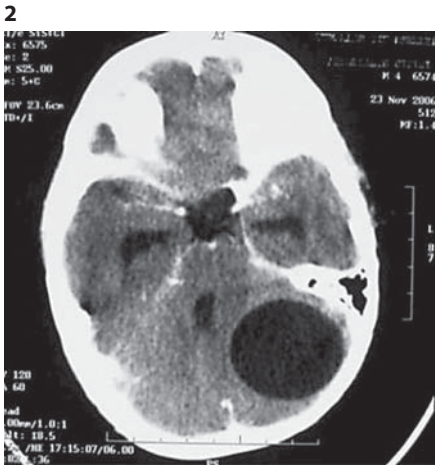
Cerebral hydatidosis is a rare disease in developed countries where it comprises about 0.05% of all intracranial mass lesions [17–21]; on the other hand, in areas endemic for the disease, hydatid cysts may account for as much as 2.5–3.5% of all intracranial mass lesions [6, 17, 22].

Fifty to 75% of intracranial hydatid cysts are seen in children. Intracranial hydatid cyst usually presents with headache, nausea and vomiting. Surgical excision is the treatment of choice. Medical therapy, such as albendazole and mebendazole, may be chosen in risky cases [23].

Radiologically, the cerebral cysts are round, thin and smooth-walled lesions that have similar density to the cerebrospinal fluid. They do not show pericystic edema unless they are infected. Pericystic contrast enhancement may be recognized due to their fibrous capsule. Hydatid cysts can be differentiated from brain abscess and cystic astrocytoma by the absence of significant rim enhancement, perifocal edema, and mural nodule [2, 3]. Other cystic lesions such as arachnoid cysts, leptomeningeal cysts, and porencephalic cysts are not spherical in shape and are not entirely surrounded by brain substance. In this study, we present 3 pediatric patients with unusually localized hydatid cysts selected among 7 cases who had been treated for primary intracranial and spinal hydatid cysts.

Patients and Methods

In this study, the patient data were reviewed in two neurosurgery departments (Departments of Neurosurgery, Faculty of Medicine, University of Harran and Mersin) between January 2001 and December 2008 to find cases who had been treated for hydatid cyst disease. Seven patients were under the age of 16 and considered as pediatric cases. Among the pediatric patients, only 3 were selected having unusually located hydatid cysts. The cysts of 2 of these patients, aged 4 and 15 years, were located in the posterior fossa and the lateral ventricle, respectively. The last case was a 9-year-old patient who had been operated on in another center for multiple cerebral cysts and multiple spinal seedings secondary to intraoperative iatrogenic cyst rupture. She was referred to our clinic due to neurologic deterioration. Spinal magnetic resonance imaging (MRI) revealed no pathologic lesions in the spinal cord; however, there were multiple cerebral cysts in the cerebrum (fig. 1a, b). Three of her cysts localized subcortically were re-



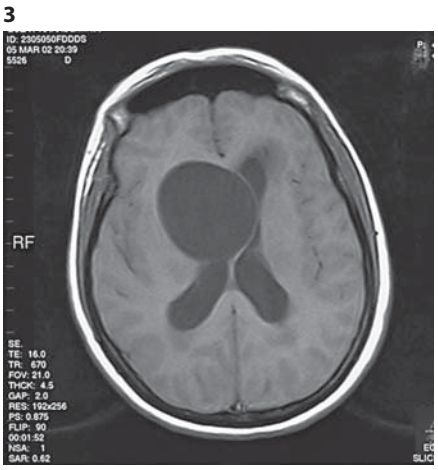


Fig. 2. Cerebellar hydatid cyst: a round, cystic lesion without perifocal edema and enhancement on CT (note the pressure effect of the lesion to the 4th ventricle).
Fig. 3. Intraventricular hydatid cyst: hypointense intraventricular lesion without gadolinium enhancement on T_1-weighted images.

moved without rupture during the operation. Six months later, an MRI scan was performed for paraplegia and urinary and bowel incontinence, showing that she had multiple spinal seedings of hydatid cysts (fig. 1c). The parents rejected a reoperation on her child. Therefore, she was put on albendazole treatment. Her course was uneventful with complete regression of the paraplegia, and an MRI scan showed that some of the cysts had disappeared and some were shrunk after 6 months (fig. 1d).

The patient who presented with a cyst located in the posterior fossa was operated immediately due to rapidly developing unconsciousness (fig. 2). A suboccipital paramedian large craniotomy and a wide cortical incision were performed; a soft catheter was introduced between the cyst and surrounding brain tissue, and saline was injected via this catheter (Dowling's technique) [24] to remove the cyst without rupture.

For the intraventricular case (fig. 3), the transcallosal approach was performed. Brain cotton was placed in the surgical field between the parenchyma and the cyst wall to prevent the contamination of the oozing cyst material. Two suction tubes were fixed near the cyst wall which was then opened by a 20-gauge cannula. The content of the cyst was controlledly aspirated with the suction tubes and the cyst wall was totally removed with a microsurgical technique using the operative microscope and the microsurgical instruments.

Discussion

The primary surgical management plan is of utmost importance for the final outcome of the patients suffering from hydatid disease, and is even more considerable for the primary hydatid cysts of the CNS.

In order to achieve the best outcome, preoperative differential diagnosis through radiological means, thorough preparation for surgery and careful follow-up of the patient are to be taken into consideration. Medical therapy is effective in selected cases with multiple cysts, and occurrence of intraoperative rupture.

The liver and the lungs are the two organs that are involved by the hydatid disease with ratios of 75 and 15%, respectively [1–3]. The involvement of all the other organs including the brain, heart, kidney, bone, breast and thyroid is 10% listed under uncommon [2] or unusual [3] localization classification.

Although cerebral hemispheric and spinal hydatid cysts are reported as unusual or uncommon cases, the deeply located ones that are seen especially in the thalamic region [13], posterior fossa [6, 25–28] and ventricles [14, 23, 29] are reported as even rarer and need greater caution. They are not just more difficult to reach, but might also cause greater morbidity due to parenchymal incision.

Cerebral hydatid cysts are usually distributed in the watershed of the middle cerebral artery [22, 30]. Erşahin et al. [6] reported that most of the cysts in their patients were localized in the supratentorial region. Our data are in accordance with the above-mentioned information with just one infratentorial involvement out of 7 cases. The majority of the cases (70%) were children in our series and this ratio also seemed to be in accordance with the related literature [6, 10, 30]. Headache, nausea and vomiting were the most common symptoms with supratentorial involvement.

Postoperative spinal seeding of hydatid cysts is an important, undesirable event. Izci et al. [31] reported 2 patients with intraoperative cyst rupture leading to spinal seeding. The history of the spinal seeding case shows some of the important features in the management plan of hydatid disease. Close follow-up of the patients with ruptured brain hydatid cysts should not be underemphasized for the spinal area that has a direct connection to the brain via cerebrospinal fluid circulation. Radiological

evaluation is to be performed periodically and should not be planned according to the presence of a neurological sign. Careful follow-up may reveal an arising lesion sooner and may give a chance for a better outcome.

Hydatid cysts of the posterior fossa are uncommon and may cause sudden deterioration of the consciousness level [6, 25–28]. Our pediatric posterior fossa case also developed this phenomenon during his physical examination. He was operated emergently and his symptoms eventually resolved. Thus, it is crucial to plan emergent surgical interventions for the cysts of this area, and this is even more important for pediatric cases in order to prevent any probable neurological deterioration.

Intraventricular involvement of hydatid cysts are also extremely rare and may be particularly hazardous [14, 23, 29]. Surgical extirpation is more difficult and prone to greater morbidity. For selected cases with cysts localized in the deeper areas of the brain, a careful attempt to collapse the cysts with a syringe and very neat microsurgical excision instead of Dowling's technique might be chosen.

All the patients were tried to be followed up periodically. The last follow-up of the posterior and intraventricular cyst patients did not reveal any recurrence at the end of the 5th and 6th year, respectively. They did not show any neurological symptoms.

The spinal seeding case with multiple cysts in the brain did not show any signs of neurological deficits either. Her MRI revealed that the size of the cysts noticed in the previous assessments diminished at the latest evaluation at the end of the 2nd year.

The preoperative diagnosis is very important in planning surgery. CT, MRI and ultrasonography are all useful techniques to localize lesions and may be used to predict the differential diagnosis of the cystic structure [32]. CT usually shows the cysts as intraparenchymal, hypodense, rounded mass lesions without perifocal edema [2, 3, 33, 34]. MRI, including diffusion and spectroscopy, precisely demonstrate location, number, cyst capsule, type of signal and enhancement, which allows diagnosis of atypical or complicated cerebral hydatid disease and appears more helpful in surgical planning [34].

The goal of surgery is to remove the cysts without rupture. Dowling's technique is very effective for total removal of the cysts without rupture [10, 24]. The cyst can be aspirated whenever Dowling's technique is not feasible, but the results from cyst aspiration are not as good as those from Dowling's technique [6]. We did not observe any recurrences in our case in whom we had performed cyst aspiration with removal of the cyst capsule.

Once a cystic lesion is found on radiological evaluation, hydatid disease must be taken into consideration in countries where hydatid disease is endemic. The differential diagnosis is even more difficult for the unusually localized cystic lesions of hydatid disease. The seriousness of the symptoms of the patients suffering from vomiting, nausea and headache may be underestimated leading to misdiagnosis. They may be treated as gastroenteritis patients, loosing time and leading to important complications. Our patients also received gastroenteritis treatment before they were admitted to our clinic. Therefore, pediatric patients mimicking gastroenteritis should be evaluated for space-occupying lesions leading to high intracranial pressure. Hydatid disease of the CNS is to be assessed in the endemic regions. It is important to remember that the cysts localized in the posterior fossa need emergent surgery.

Spinal seeding is to be taken into consideration for cases with intraoperative intracranial cyst rupture and appropriate radiological evaluation should be planned in the follow-up period.

To conclude, hydatid cyst cases, although they are very rare, should be taken into consideration for the abovementioned conditions and great caution is to be taken to prompt diagnosis and appropriate therapy.

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