

Large primary cerebral hydatid cysts in children

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ABSTRACT

Hydatid cyst disease in childhood is still a serious health problem in the rural areas of Turkey and other places where the parasite is endemic. The brain involvement rate varies from 1-2% in hydatid disease. Especially in children, hydatid cysts can grow to enormous sizes, and the cases can remain neurologically intact. In this report, a 7-year-old boy, a 15-year-old girl, and a 10-year-old girl, in whom large primary brain hydatid cysts were diagnosed radiologically and treated surgically, are presented. Severe headache in childhood should be taken into consideration carefully in countries where hydatid disease is seen.

Neurosciences 2006; Vol. 11 (4): 318-321

Hydatid cyst disease is still a serious health problem in the rural areas of Turkey and other places where the parasite is endemic.¹⁻⁴ It is hyper endemic in Iran, Iraq, Jordan, Morocco, Libya, Tunisia, and Algeria, and endemic in Egypt.⁵ In Tunisia, the annual human incidence of surgical cases of cystic echinococcosis is approximately 15 cases/100,000.³ The incidence of surgically confirmed cystic echinococcosis in eastern Libya was estimated to be at least 4.2 cases/100,000, with significantly more female cases than male. Ultrasound examinations and serology revealed that 1.6% of those living in northern Libya had the disease.⁶ Cerebral hydatid disease is very rare and brain involvement in hydatid disease occurs in 1-2% of all *Echinococcus granulosus* infections.^{1,2,7,8} Fifty to 75% of intracranial hydatid cysts are seen in children;² where they can grow to enormous sizes and remain neurologically intact.⁸ In this article, we present 3 children harboring primary cerebral hydatid cysts, who were surgically treated; the relevant literature is discussed.

Case Reports. Patient 1. A 7-year-old boy was admitted to our Neurosurgical Clinic with headache and hypoesthesia of the left arm for 15 days. Neurological examination was normal. Liver function, abdominal ultrasonography and the other laboratory tests were normal. Chest x-ray and echocardiography also revealed no pathologic findings. A CT scan and MRI of the brain showed a large unilocular, round, without rim enhancement, cystic space-occupying lesion in the right parietal region approximately 8 x 8 x 7 cm in diameter with midline shift to the left hemisphere. No peripheral edema was found around the cyst (**Figure 1a**). A large right parietal craniotomy was performed to clear the right parietal region. A soft rubber catheter was inserted between the hydatid cyst and surrounding brain tissue, and warm saline was injected through this catheter (Dowling's technique). The cyst was totally removed unruptured. His postoperative course was uneventful. He was treated with 10 mg/kg of albendazole 3 times per day for 4 months. A 6-month MRI follow-up showed no relapse, and the brain was normal (**Figure 1b**).

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Received 21st December 2005. Accepted for publication in final form 22nd March 2006.

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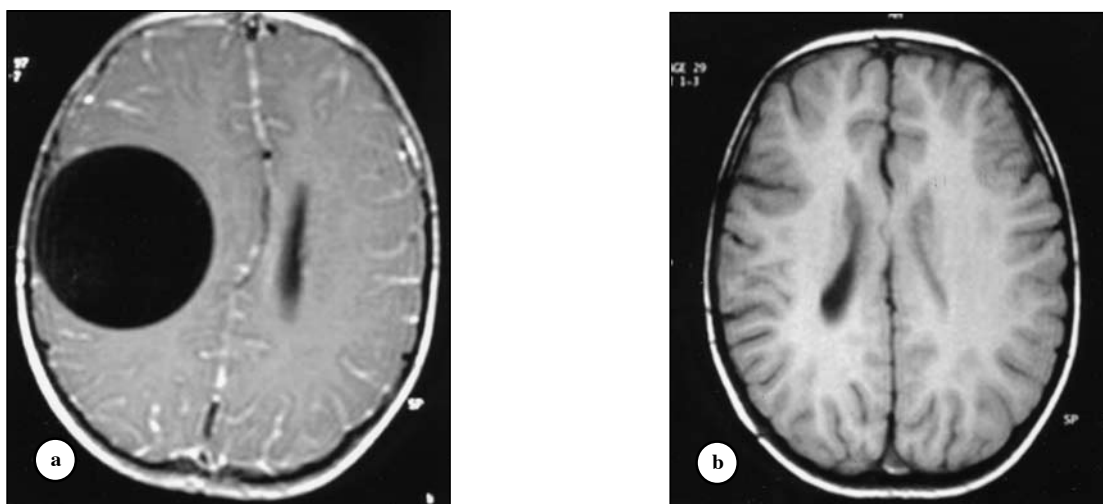


Figure 1 - Magnetic resonance images showing **a)** right parietal large unilocular, cystic lesion, with midline shift. There is no rim enhancement, nor surrounding brain edema. **b)** The follow-up MRI shows complete resolution of the cyst.



Figure 2 - A CT scan showing a large unilocular, cystic lesion located in the left fronto-parietal region with displacement of the ventricular system to the opposite side.

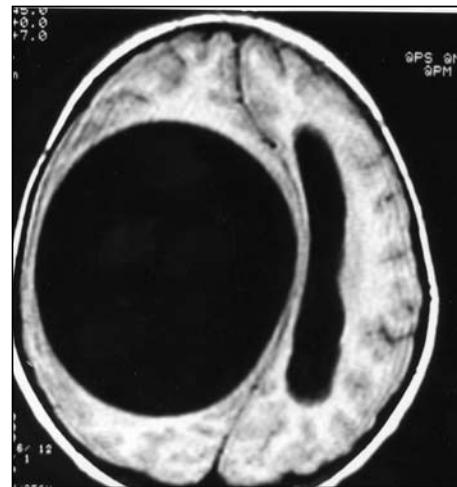


Figure 3 - Axial MRI revealing a large unilocular, cystic lesion located in the right parieto-occipital region with midline shift to the left hemisphere, without rim enhancement. Edema is not seen in the surrounding brain tissue.

Patient 2. A previously healthy 15-year-old girl was referred to our Neurosurgical Clinic complaining of headache and vomiting for one month's duration. Neurological examination did not reveal any significant abnormality. A CT scan of the brain showed a large unilocular, cystic lesion with clearly defined borders, in the left fronto-parietal region (approximately 9 x 8 cm in diameter). There was marked compression of the left cerebral hemisphere and displacement of the ventricular system to the opposite side. The margins were smooth; the CT density of the cystic fluid and its absorption value were for cerebrospinal fluid (CSF). No peripheral edema and rim enhancement

were noted around the cyst (**Figure 2**). Liver function, serological tests, and abdominal ultrasonographic examination were within normal limits. Chest x-ray and echocardiography also revealed no pathologic findings. A large left frontoparietal bone flap was turned. The hydatid cyst spontaneously ruptured when the duramater was opened. The fluid of the cyst was evacuated with suction, and wall of the cyst was totally removed. The cavity of the cyst and surgical field was irrigated with warm 3% NaCl for 10 minutes. The early postoperative course was uneventful. She was treated with 10 mg/kg of albendazole 3 times per day for 6 months. The postoperative control CT scan

showed fluid for CSF in the surgical area. She has been monitored for 15 months and has not developed any neurological deficit, and is without occurrence of any problems.

Patient 3. A previously healthy 10-year-old girl was referred to our Neurosurgical Clinic, complaining of headache, vomiting and weakness of the left side for 3 weeks. Neurological examination showed left hemiparesis (strength of muscle was 3/5). ACT scan of the brain showed a large unilocular, cystic lesion with clearly defined borders, in the right fronto-temporo-parieto-occipital regions (approximately 10 x 11 cm in diameter). There was marked compression of the right cerebral hemisphere and displacement of the ventricular system to the opposite side. The margins were smooth; the CT density of the cystic fluid and its absorption value were for CSF. No peripheral edema and rim enhancement were noted around the cyst. An MRI of the brain showed a large unilocular, round, cystic space-occupying lesion in the right fronto-temporo-parieto-occipital regions approximately 10 x 11 x 10 cm in diameter with midline shift to the left hemisphere, without rim enhancement. No peripheral edema was found around the cyst (**Figure 3**). Liver function, serological tests, and abdominal ultrasonographic examination were within normal limits. Chest x-ray and echocardiography also revealed no pathologic findings. A large right fronto-parieto-occipital bone flap was turned. A soft rubber catheter was inserted between the hydatid cyst and surrounding brain tissue, and warm saline was injected through this catheter (Dowling's technique). The cyst was totally removed unruptured. Her postoperative course was uneventful. She was treated with 10 mg/kg of albendazole 3 times per day.

Discussion. Cerebral hydatid disease is very rare, and especially in children, hydatid cysts can grow to enormous sizes, taking advantage of the elastic bony structure, open sutures, and higher compressibility of the neural tissue.⁸ In this age group, central nervous system (CNS) hydatidosis can be associated with involvement of other organs such as liver and lung, or may be an isolated infestation of the CNS and its coverings. Only embryos that succeed in passing through the filtering barrier systems in the liver and lung reach the brain or spinal cord via the systemic circulation. Basically, the cystic lesions can be single (77%) or multiple (23%).⁴ Multiple larval intake and spontaneous, traumatic or surgical rupture of a brain hydatid cyst may cause multiple intracranial hydatidosis.³ Primary hydatid cysts are usually solitary, and secondary cysts are usually multiple. The cysts enlarge slowly, rates of approximately 1 cm

per year are quoted, but this is variable and may be higher in children.² Several observations have been made about the growth rate of hydatid cysts. Altinors et al¹ reported a growth rate of 7 mm/month.

The development of symptoms is slow, neurological deficits appear late and are often preceded by signs of increased intracranial pressure.^{1-4,9} Other symptoms, such as weakness in the limbs and gait disorders, may vary with the location of the cyst. Papilledema is usually present in patients with intracranial hydatid cyst at the time of diagnosis.^{2,4} In children and adolescents, the clinical picture usually includes the cardinal symptoms of increased intracranial pressure, whereas focal findings such as hemiparesis, speech disorders, and hemianopsia, sometimes associated with epileptic seizures, are more prevalent in the older age group.^{2,4} In our cases, we detected similar symptoms.

The primary hydatid cysts, are frequently found in the cerebral hemisphere and generally cause significant ventricular distortion and midline shift, as in the present cases. They are distributed in the watershed area of the middle cerebral artery, particularly in the parietal and frontal lobe.^{1,4} Because of their large sizes, they usually lie only a few millimeters below the cortex, which is flattened over them and appears bluish, as in our second and third cases. Their outer surface has a characteristic pearly white or grayish color.^{1,2}

Hydatid cysts are usually acquired in childhood and grow slowly but progressively. The patient presenting with this condition is most commonly a child or young adult. The correct preoperative diagnosis, formerly arduous, can now be established by CT and MRI.^{1,4,7} The CT and MRI not only have tremendously increased diagnostic specificity, but also allowed us to visualize the outermost margin of the cyst, thus helping the surgeon to plan the cortical incision and accurately approach the lesion.^{1,4,7} In principle, preoperative diagnosis of hydatid disease is essential, because rupture and dissemination of the cyst may result in recurrence and even death.⁴

The aim of surgery is to remove the cysts in total without rupture. Dowling's technique is very effective for total extirpation of the cysts.^{1,4} Location and multiplicity are the 2 main problems faced by surgeons in treating this disease. Lesions at deep locations, cysts in eloquent areas, and sites bounded by bone tissue pose barriers to total removal of cysts. In addition to the problems that are associated with their growth, hydatid cysts can rupture either spontaneously, as a result of trauma, or during surgery.^{1,3} This may result in an anaphylactic reaction with circulatory collapse and cardiac arrest.^{1,3,4}

The intraoperative rupture rates for intracranial hydatid cysts were 16.9% and 25.6% in the cooperative study and in the literature survey conducted by Altinors et al.¹ The accepted procedure in cases of intraoperative rupture is aspiration of the cyst contents, extirpation of the cyst wall, and as a precaution, extensive irrigation of the surgical field with hypertonic solution to prevent recurrence.^{1-4,9}

The results of surgical treatment depend on several factors, including the location, size, and multiplicity of the cysts. It is considered that patients with multiple cysts have high morbidity and mortality rates due to the number of surgical interventions needed to remove all their cysts, but these patients frequently tolerate multiple surgical procedures. In the postoperative period, subdural effusions and pencephalic cysts can occur as a complication in children and adolescents.^{1,4,9} The shunting procedure is a technique used when postoperative CT/MRI shows ventricular dilatation, subdural effusion, or pencephalic cyst, or both.^{1,4,9} The reported recurrence rate for intracranial hydatid cysts ranges from 14-25%,^{4,9} whereas the reported mortality rate is 10%.⁴

Chemotherapy may be the initial form of treatment for small multiple cysts located in eloquent areas of the brain in patients with no or minimal neurological deficits.¹ Supplementary chemotherapy can be used such as mebendazole or albendazole.^{1,2,10} A slightly greater efficacy as related to rates of complete cure and improvement has been obtained with albendazole. In cases of cerebral echinococcosis, there are single reports of successful treatment using a course of albendazole alone, without surgery.¹⁰ No agreement has been reached concerning the duration of chemotherapy, but most authors recommended a treatment course lasting 4 months.¹⁰

In conclusion, hydatid cyst disease in childhood is still a serious health problem especially in the rural areas. In children, cysts can grow to enormous sizes, and the cases can remain neurologically intact. For this reason, severe headache in childhood should be taken into consideration carefully in countries where hydatid disease is seen.

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