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ORIGINAL PAPER

Intracranial hydatid cyst in children: report of 30 cases

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Abstract
Purpose To analyze the clinical manifestations, radiological features, and surgical outcomes in 30 pediatric intracranial hydatid cysts.
Methods We reviewed the clinical, radiological, and surgical aspects of pediatric intracranial hydatid cysts patients who received surgical treatment at the Neurosurgical department of Xinjiang Medical University between the years 1985 to 2007, retrospectively.
Results Headache and vomiting were the most common initial symptoms in our series. Neurological deficits from the mass effect of the cysts were seen in 15 cases, including hemiparesis, visual deficit, and diplopia. Epilepsy occurred only in one patient with temporal lobe hydatid cyst. On computed tomography (CT), it presented as a round-shaped and thin-walled homogeneous low-density cystic lesion without surrounding edema and enhancement. Only five patients had a magnetic resonance imaging (MRI) scan, and presented low signal intensity on T1-weighted image and high signal intensity on T2-weighted image. Surgical removal of cyst was performed in all cases and intact removal was done in 29 cases. However, one cyst ruptured during the dissection of cyst wall, thus, resulting in one death. There were no additional neurological deficits which were caused directly by surgery.
Conclusion Increased intracranial pressure is common in patients with cerebral hydatid disease. CT and MRI are the first-line diagnostic procedures. Surgery is the treatment of choice for the majority of intracranial hydatid cysts. Multiple and deep seated lesions should receive medical treatment postoperatively.

Keywords Intracranial hydatid cysts · Children · Treatment · Surgery

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Introduction

Hydatid disease is caused by *Echinococcus granulosus* or *Echinococcus multilocularis* during the larval stage of the tapeworms. It occurs most commonly in the liver and lung. Intracranial hydatid cysts are less common, which only accounts for 0.5% to 3% of all hydatid diseases [7, 8, 10]. Humans can play a role as an intermediate hosts in the tapeworm life cycle. Humans become infected by ingesting tapeworm eggs passed from an infected carnivore, which most frequently happens when individuals handle or have a contact with infected carnivores or inadvertently ingest food or drink which is contaminated with fecal material containing tapeworm eggs [1, 10]. Today, there are no exact national or international figures on how many people have hydatid cyst. Hydatid cysts are endemic in many countries, including China, especially in Xinjiang Province. Since 1985, we have treated 56 intracranial hydatid cysts at our department, most of whom were children, especially from the Kazakh ethnic group. In this article, we report on the clinical manifestations, radiological features, and surgical outcomes in 30 pediatric cases with intracranial hydatid cysts that underwent surgery at the Neurosurgery Department at Xinjiang Medical University since 1985.

Patients and methods

We reviewed all the clinical, radiological, and immunological data of all patients who were diagnosed as harboring an intracranial hydatid cyst since 1985, retrospectively. There were 30 children found to have an intracranial hydatid cyst who subsequently underwent surgery at our department. The patients' population consisted of 21 male and nine female patients, ages ranging from 4–18 years, with median age being 11 years. There were 12 Kazakh, eight Chinese, six Uyghur, and four Mongol patients. All patients were given a head computed tomography (CT) scan or magnetic resonance imaging (MRI), chest X-ray, abdominal ultrasound, and immunological test preoperatively, and head CT scan or MRI 1 week and annually after that. All surgeries were performed under general anesthesia. Craniotomies were performed big enough to facilitate the removal of the hydatid cyst intact. Dowling's technique was used in all cases to dissect the cyst wall from the surrounding brain parenchyma with irrigation. Hypertonic saline irrigation of the cystic cavity (20%) was used for ruptured cysts intraoperatively. Albendazole was given to patients who had multiple organ involvement, or multiple cysts inside the cranial cavity and still have residual cysts after surgery, and cyst which was ruptured intraoperatively. The total daily dose was calculated as 15 mg/kg.d, administered twice a day orally for 6 months postoperatively. Follow-up times ranged from 2 months to 20 years with the mean follow-up time being 4.5 years.

Results

Fifty-six patients with intracranial hydatid cysts received treatment at our department since 1985. Among them, 30 children were found to have an intracranial hydatid cyst and underwent surgery. Headache and vomiting were the most common chief complaints in 26 of these patients (87%) followed by hemiparesis in nine patients. Less common complaints included decreased visual acuity and visual deficit in five patients, cerebellar ataxia in four patients, subcutaneous mass in two patients, aphasia in one patient, coma in one patient, epilepsy in one patient, and ptosis in one patient. Symptom duration time ranged from 2 weeks to 6 years. All patients had a head CT scan, and found to have a cystic, well-demarcated, thin-smoothed walled lesion without rim enhancement, except one patient who presented as an asymptomatic right occipital subcutaneous mass manifested as a heterodensity mass lesion with displacement of right cerebellar hemisphere, erosion of occipital bone, and obstructive hydrocephalus on non-contrasted CT scan. Only five patients had MRI scan and presented as low signal intensity on T1-weighted image and high signal intensity on T2-weighted image. Eight patients took an immunological test. Casoni reaction was tested in eight patients with positive results in five patients. All patients underwent surgery. Only one cyst was ruptured during the operation although the residual cavity was irrigated with 20% saline, but the patient developed severe allergic shock from the leakage of cystic contents and died on the second postoperative day. Only three patients had multiple cysts, among them one patient who had five cysts (Fig. 1), another had three cysts, and the other one had two cysts. The patient who had two cysts had them punctured at a local hospital, after that she developed two intracranial cysts and was transferred to our hospital for the removal of cysts. The cysts were removed without rupture, but it reoccurred after 8 months, which lead to the patient's death. Seven patients had coexisting liver cysts; one patient has multiple organ involvement with liver and lung cysts. A total 39 intracranial cysts were removed surgically using Dowling's technique. Follow-up time range from 2 months to 20 years. Mean follow-up time was 4.5 years. There were no recurrences among the patients with solitary lesions which were removed intact (Fig. 2). Among the patients with multiple cysts, two of them had reoccurring cysts. Five out of eight patients with multiple organ involvement did not have any reoccurrence after the removal of co-existing liver or/with lung cysts. Albendazole was given to patients who had multiple organ involvement or multiple cysts inside the cranial cavity and still have residual cysts after surgery and cyst which was ruptured intraoperatively. The daily total dose was calculated as 15 mg/kg.d, administered twice a day orally for

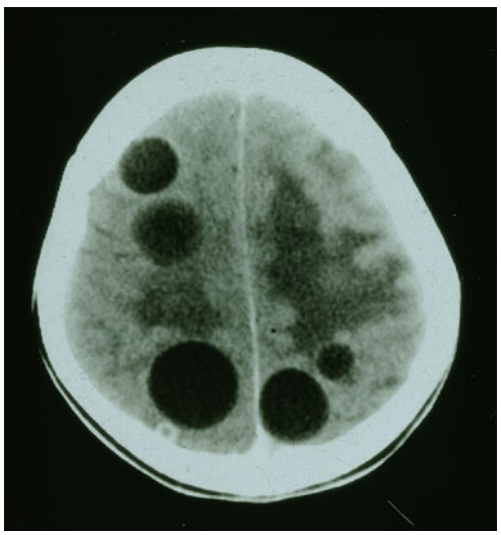

Fig. 1 Pre-operative CT scan of a 6-year-old Kazakh boy with increased intracranial pressure and hemiparesis which showed five intracranial cystic masses on frontal and parietal regions, manifested as thin-walled, well-demarcated, and round cystic lesions subsequently diagnosed as hydatid cysts intraoperatively

Fig. 2 Unruptured hydatid cysts which were delivered from intracranial space using Dowling's technique

6 months postoperatively. Clinical symptoms were improved dramatically in all patients after the removal of the cyst, except in one patient who had a long medical history with occipital cyst still has visual problems after follow-up to 4 years (Fig. 3a-c), one patient with multiple reoccurring cysts had died 8 months after the first operation. Patients' demographics, the cysts' locations, outcome, preoperative/postoperative neurologic deficits, mortality/morbidity rates, and the volume of each individual cyst are listed in Table 1.

Discussion

Echinococcosis, also known as hydatid disease, hydatid cyst, unilocular hydatid disease, or cystic echinococcosis is a potentially fatal parasitic disease that can affect many animals, including wildlife, commercial livestock, and humans. The disease results from infection by tapeworm larvae of the genus Echinococcus—notably *E. granulosus*, *E. multilocularis*, *Echinococcus vogeli* and *Echinococcus oligarthrus*. Like many other parasite infections, the course of echinococcosis infection is complex. The worm has a lifecycle that requires definitive hosts and intermediate hosts. Definitive hosts are normally carnivores such as dogs, while intermediate hosts are usually herbivores such as sheep and cattle. Humans also function as intermediate

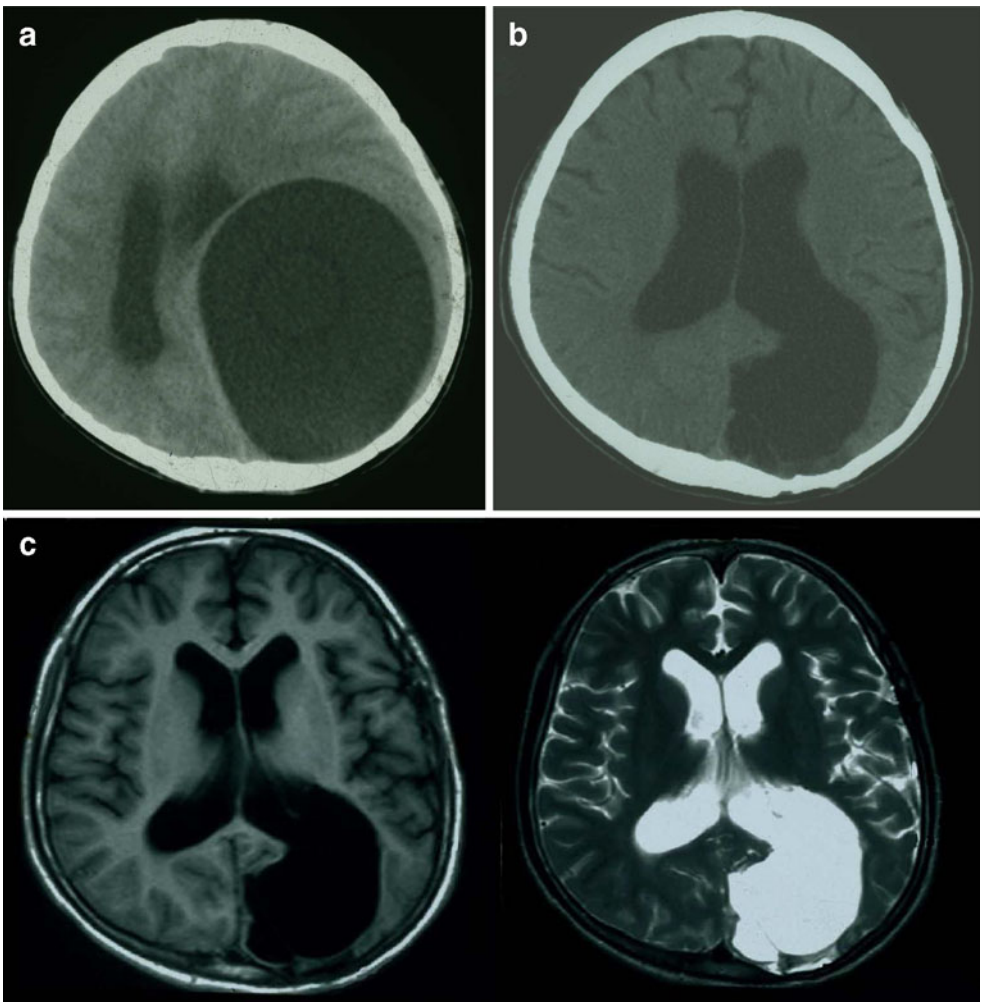

Fig. 3 **a** CT scan of a seven-year-old boy with increased intracranial pressure and visual disorder which showed a giant left temporo-occipital hydatid cyst with significant midline shift. **b** Post-operative CT scan of the boy showed left occipital pore-encephaly after the removal of the cyst. **c** Post-operative MRI scans of the boy showed left occipital pore-encephaly after the removal of the temporo-occipital hydatid cyst

Table 1 Summary of 30 pediatric cases with intracranial hydatid cysts

Patient	Sex	Nationality	Age	Preoperative neurological deficit	Cyst location	Volume(mm^3)	Postoperative deficit	Morbidity	Mortality	Outcome
1	Male	Kazakh	11	Left hemiparesis	Frontoprietal	61×70×60	No	0	0	Excellent
2	Female	Kazakh	10	Ataxia, decreased visual acuity	Cerebellar vermis	47×46×40	No	0	0	Excellent
3	Male	Uyghur	13	No	Occipital extradural	83.6×147×60	No	0	0	Excellent
4	Male	Chinese	11	No	Frontal	38×55×30	No	0	0	Excellent
5	Male	Uyghur	18	No	Temporal	63×65×60	No	0	0	Excellent
6	Female	Mongol	13	Coma	Occipitoprietal	68×54×70	No	0		Excellent
7	Male	Chinese	4	Epilepsy	Temporal	86×74×90	No	0	0	Excellent
8	Female	Kazakh	5	Hemiparesis	Frontoprietal	94.5×81.3×80	No	0	0	Excellent
9	Female	Uyghur	6	Hemiparesis	Temporopraietal	84×87.1×70	No			Excellent
10	Male	Kazakh	6	Hemiparesis	Frontotemporoprietal (multiple)	103×86×90	No	Re-occured	0	Good
11	Male	Chinese	6	Ataxia	Fourth ventricle	39×39×40	No	0	0	Excellent
12	Male	Mongol	5	Decreased visual acuity	Parietotemporo-occipital (multiple)	170×150×50	No	Re-occured	1	Death
13	Male	Kazakh	10	Hemiparesis	Frontotemporoprietal	116×80×80	No	0	0	Excellent
14	Male	Uyghur	5	No	Temporal	27×30×40	No	0	0	Excellent
15	Female	Kazakh	8	Hemiparesis, aphasia	Frontotemporoprietal	100×80×80	No	0	0	Excellent
16	Male	Chinese	8	Ataxia	Fourth Ventricle	29×28×10	Coma	Coma	1	Death
17	Male	Kazakh	10	Decreased visual acuity	Parietotemporo-occipital (multiple)	95×82×80	No	0	0	Excellent
18	Female	Kazakh	7	Parietal subcutaneous mass	Parieto-occipital	70×60×80	No	0	0	Excellent
19	Male	Chinese	12	No	Frontal	80×20×100	No	0		Excellent
20	Male	Kazakh	7	Visual deficit	Temporo-occipital	58×65×60	Visual deficit	Visual deficit	0	Good
21	Female	Chinese	7	No	Frontotemporal	120×80×40	No	0		Excellent
22	Female	Kazakh	12	Hemiparesis	Frontoprietal	67×63×70	No	0	0	Excellent
23	Male	Kazakh	7	Ataxia	Cerebellar vermis	47×48×50	No	0	0	Excellent
24	Male	Chinese	6	No	Temporal	30×28×40	No	0		Excellent
25	Male	Kazakh	15	Ptosis	Parasellar	43×41×40	No	0	0	Excellent
26	Male	Chinese	6	Hemiparesis	Frontoprietal	52×60×50	No	0	0	Excellent
27	Female	Mongol	10	No	Frontotemporal	15×15×15	No	0	0	Excellent
28	Male	Mongol	7	Decreased visual acuity	Ocipitoprietal	103×86×90	No	0	0	Excellent
29	Male	Uyghur	10	No	Frontal	50×54×50	No	0	0	Excellent
30	Male	Uyghur	11	Hemiparesis	Frontoprietal	81×81×80	No	0	0	Excellent

hosts, although they are usually a "dead end" for the parasitic infection cycle. The disease cycle begins with an adult tapeworm infecting the intestinal tract of the definitive host [1, 10, 12]. The adult tapeworm then produces eggs which are expelled in the host's feces. Intermediate hosts become infected by ingesting the eggs of the parasite. Inside the intermediate host, the eggs hatch and release tiny hooked embryos which travel in the bloodstream, eventually lodging in an organ such as the liver, lungs, brain, and kidneys, etc. There, they develop into hydatid cysts. Inside these cysts grow thousands of tapeworm larvae, the next stage in the life cycle of the parasite. When the intermediate host is predated or scavenged by the definitive host, the larvae are eaten and develop into adult tapeworms, and the infection cycle restarts. Echinococcosis infection causes large cysts to develop in intermediate hosts [1, 4, 7]. Symptoms arise as the cysts grow bigger and start eroding and/or putting pressure on blood vessels and organs. Large cysts can also cause shock if they happen to rupture. Infection with *E. granulosus*, although common in Mediterranean countries, is also common in Northwest China, especially in Xinjiang Uyghur Autonomous Region.

Xinjiang Uyghur Autonoumus Region is known as an endemic area not only for cystic echinococcosis, but also for alveolar echinococcosis. In this highly endemic area, the domestic dog acts as the main definitive host of *E. granulosus* with an infection rate between 7% and 71%, while sheep acting as the main intermediate host with an infection rate varying between 3.3% and 90% [3]. There are about 19 million people in Xinjiang Uyghur Autonoumus Region, which consists of 13 nationalities, among them Uygur, Kazakh, Mongolian, and Chinese are predominant. We have treated 12 Kazakh (40%), eight Uyghur (27%), six Chinese (20%), and four Mongol patients (13%). Although Kazakhs only consist 8% of the population in Xinjiang, we have more Kazakh patients compared to other nationalities. It is considered that echinococcosis is a regional disease and all of our patients are from the endemic areas where these nationalities live together. Transmission to humans, however, is a complex problem. Kazakhs and Mongols are still practicing nomadic life and living on raising livestock such as sheep, goats, cows, horses, and camels. Being nomads, they exclusively raise a dog which acts as a protector of their family and livestock from danger, the dog is also looked at as a family member. Thus, they have close contact with their livestock and dogs in their everyday life. Mutton is the favorite meat of all nationalities in our region it is especially the main dish of the Kazakhs and Mongols. Home slaughtering is still practiced widely in our region. Usually Kazakh and Mongols feed the dogs with the internal organs which are suspected to have infections which include echinococcosis infection; the common practices of home slaughter and of feeding dogs on offal containing hydatid cysts facilitate the life cycle of the parasite. The feces of the infected dog contaminate the grass land, water, etc. and cause to spread and transmission of the disease. Direct contact of children with dogs and ingestion of water, vegetables, and foods contaminated by worm eggs are the chief mode of transmission for human hydatidosis [5]. Nowadays, livestock owners are taking care of the prevention and treatment of echinococcosis among their livestock. Nomads typically use antithelmintic drugs to treat their animals to prevent or treat echinococcus infection, but less educated nomads do not know how to treat the abnormal-looking internal organs after home slaughtering. Usually, they use antithelmintic drugs only for live stocks such as sheep, goats, etc., but not for dogs. Also, nomadic life is still different from modern life where tap water, kindergarten, toilets, and shower are not available, and they still practicing uninspected home slaughtering, so poor personal and general sanitation also contribute to the transmission and spread of the echinococcosis. On the basis of hydatid control efforts for several years, the Ministry of Public Health of China established some means to control echinococcosis which include extensive health education, sanitation of slaughtering, and management and deworming of dogs. As a result, we have fewer patients with hydated cyst in the last decade.

Intracranial hydatid cyst presents with a wide range of clinical manifestations which mainly depends on the location and size of a given hydatid cyst [1]. It is a relatively slow growing benign lesion. The annual growth rate was reported to be in the range of 1–10 cm [6]. According to our review, supratentorial hydatid cysts tended to be larger than the inferiortentorial counterparts when causing symptoms. Duration time of symptom is found to be an independent predictor of the outcome. Most common symptoms are headache and vomiting, followed by weakness of extremities. Also these symptoms tend to be relieved after the successful removal of cysts. However, compared to other intracranial mass lesions, it can cause relatively few neurological deficits, which can be improved dramatically after successful removal of the cyst. According to the literature, multiple organ involvement is not rare in hydatid disease, but we have fewer patients with multiple organ involvement in our series compared to other clinical reports, but the exact epidemiology is unclear.

Skull radiographs show the effects of increased intracranial pressure such as thinning of the vault and diastases of sutures in children. In the pre-CT era, ventriculography and angiography were used in central nervous system hydatidosis with low diagnostic sensitivity and specificity. At that time, angiography replaced air studies in the investigation of intracranial lesions. With the clinical availability of CT and MRI, it has become easy to diagnose

an intracranial hydatid cyst, especially in endemic areas. CT and MRI can not only clearly show us the exact location of a cyst, but also provide us with more information such as the number of cysts, daughter cyst, and infected cyst, etc. Single lesions presents as a thin-walled, well-demarcated, and round cystic lesion on CT. Contents of the cyst manifest as hypo-signal intensity on T1-weighted images and hyper-signal intensity on T2-weighted images, and cyst wall presents hypo-signal intensity on both T1-weighted and T2-weighted MRI images [2, 9]. Almost all hydatid cysts manifest typical appearance on CT and MRI in our series. Except for one patient who had an occipital extradural mass, manifested as a mixed density, bone erosion with marked ring enhancement on contrast CT scan, and subsequently diagnosed as an infected hydatid cyst after pathological examination. There is no enhancement and surrounding edema of hydatid cyst in non-infected cysts. Differential diagnosis of a hydatid cyst against other cystic lesions, such as arachnoid cyst, porencephalic cyst, and cystic tumors is not difficult when the imaging features of these lesions are well-known to both radiologists and neurosurgeons.

Serological tests are important while diagnosing and post-operative follow-up of a hydatid cyst, but the exact value of such kind of tests is limited because of their low sensitivity [1, 10]. With the advent of CT and MRI, making the diagnosis of the intracranial hydatid cyst became easier. According to our experience, old serological methods were low in sensitivity and specificity. It is always negative in intracranial solitary lesions, but more sensitive in diagnosing cysts with multiple organs involvement especially liver. Recently available immunological methods such as enzyme-linked immunosorbent assay seem to have more clinical value.

Treatment methods of intracranial hydatid cysts include surgical removal, aspiration, and chemotherapy. Surgical removal is the treatment of choice of all intracranial hydatid cysts in children whenever possible. Surgery of solitary intracranial hydatid cyst is relatively easy with very promising outcome if the cyst is removed intact because rupture and dissemination of the cyst may result in recurrence or even death. The Dowling–Orlando technique is very useful during the removal of intraparanchymal cysts, which requires a wide osteoplastic flap, exposure of the cyst by a wide incision, an inclined position of the head, and saline irrigation by means of a rubber catheter. Aspiration of the cystic contents through puncturing during the surgery is another alternative method for the treatment of deep-seated hydatid cysts, but the rate rupture and reoccurrence is high, so we do not recommend the application of this technique.

Surgery-related complications of hydatid cysts are rare, which include subdural effusions and porencephalic cysts, hemorrhage in the residual cavity, and hydrocephalus in children may be treated conservatively. Occasionally, shunting is necessary for subdural effusions and hydrocephalus [2, 10].

Chemotherapy is being an adjunct treatment for patients with inoperable multiple cysts or deep-seated cysts and in those who are not good candidates for surgery or may be suffering from recurrent cysts [13]. Reported to be clinically effective, antihelminthic agents for the medical treatment of hydatid cyst are Mebendazole and Albendazole [11], but its effectiveness is still unclear. In our experiences, these drugs were not shown to be as effective as shown in some literature. In our series, Albendazole was given to patients who had multiple organ involvement or multiple cysts inside the cranial cavity and still have residual cysts after surgery and cyst which was ruptured intraoperatively. We do not recommend to routinely use Albendazole as a preventive drug after the intact removal of intracranial hydatid cyst or cysts. We need to do further randomized clinical trials to test the pursued effectiveness of these drugs and developing new drugs.

Conclusion

Increased intracranial pressure is common in patients with cerebral hydatid disease. CT and MRI are the first-line diagnostic procedures. Surgery is the treatment of choice for the majority of intracranial hydatid cyst. Multiple lesions should receive medical treatment postoperatively.

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