

CLINICORADIOLOGICAL FINDINGS AND TREATMENT OUTCOME IN PATIENTS WITH INTRACRANIAL HYDATID CYST

Z. Miabi^{*1}, H. Hashemi², M. Ghaffarpour³, H. Ghelichnia³ and R. Media⁴

1) Department of Radiology, School of Medicine, Tabriz University of Medical Sciences, Tabriz, Iran

2) Department of Radiology, Imam Khomeini Hospital, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

3) Department of Neurology, Imam Khomeini Hospital, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

4) Department of Radiology, QEII Health Sciences Center, Dalhousie University Halifax, Nova Scotia Canada

Abstract- Human infection with *echinococcus granulosus* is a common disease throughout south America, The Mediterranean littoral, The Middle East, Central Asia and East Africa, which usually occur in children and young adults. Formation of avascular cystic lesions in the liver, kidney, pancreas, bones, vitreous and brain can cause protean of signs and symptoms. Intracranial cysts usually present with focal neurological deficit and features of raised intracranial pressure. Primary hydatid disease of the brain is a rare entity but may pose various diagnostic problems. In this study we report the clinicoradiological findings, treatment outcome and some other properties of intracranial hydatid cysts in 24 cases, emphasizing the fact that hydatid cyst should always be suspected in cystic lesions affecting intracranial cavity specially in endemic areas. Sixty five percent of our patients were children and young adults, 85% of whom came from rural areas. We found in contrast to other studies a female predominance (58%). Headache and vomiting were the most common symptoms. All but one of the patients had a solitary lesion in the cerebral hemisphere. In 21/24 (87%), cysts were removed intact. Four patients (three ruptured cysts during surgery and one case with additional cyst in the lung) received mebendazole (800 mg daily). Surgical mortality and postoperative complications were 8.3 and 20.8% respectively.

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INTRODUCTION

Echinococcosis, known also as hydatidosis or hydatid disease, is a zoonosis in which the definitive host is a carnivore that harbors the adult tapeworm in the small intestine.

The carnivore becomes infected by ingesting the larval form in the tissue of the intermediate host. The intermediate host, chiefly herbivorous animals but also humans, become infected by ingestion of tapeworm eggs, passed in carnivore feces.

Hydatid disease is usually transferred to adults by uncooked foods, whereas in children infection commonly takes place via accidental contamination by direct contact with feces of dogs. The larvae reach the brain after passing through the liver and lung filters. In rare instances intracranial cysts are

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*** Corresponding Author:**

Z. Miabi, Department of Radiology, School of Medicine, Tabriz University of Medical Sciences, Tabriz, Iran
Tel: +98 411 4790364, 3346960, Fax: +98 411 4796969, 3356612
E-mail: miabiz@tbzmed.ac.ir

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caused by embolism of cardiac echinococcosis (1). Many authors have reported different growth rates (1 to 5 cm per year) for hydatid cysts, which probably is higher in children.

Hydatidosis is an important public health problem in many parts of the world especially in rural areas, where sheep and cattle are raised (2, 3). Endemic foci are in Eastern Europe, the Mediterranean countries, Australia, New Zealand, India, Russia and south Africa (4, 5). Involvement of the central nervous system (CNS) is seen only in 2-3% of patients and mainly affects cerebral hemispheres but infection may be located in the extradural space, cavernous sinus, intradiploic or eyeball (6-10). Hydatid cysts have three layers except for those of the bone, which do not have outer or host layer (11).

The immunoblot test, where available, is the test of choice (98% specific and 91% sensitive). The arc 5 test is also diagnostic except for cross reactions with *Taenia solium* cysticercosis infection. Several other tests (enzyme-linked-immunosorbent-assay [ELISA] and indirect hemagglutination and immunofluorescence) are useful but both false-negative and false-positive results are common. The Casoni skin test has been abandoned because of poor specificity (11). Hydatid cysts should be differentiated from malignant or non-malignant cysts and abscesses.

Definitive treatment still consists of surgical removal. Decision making to choose between the surgery and medical treatment with albendazole or mebendazole must take into account (a) surgical mortality ($\geq 2\%$), postoperative complications (10-25%) and recurrence rate after surgery (2-25%); and (b) cure rate with medical treatment (30-40%). One approach is to give a course of treatment to selected asymptomatic patients whose cysts are small and are not in danger of rupture. If after 6-9 months the cyst has not disappeared or clearly died, it can then be removed surgically.

In this study, we report the clinicoradiological findings, treatment outcome and some other properties of intracranial hydatid cysts in 24 cases with intracranial cystic lesion confirmed to be hydatid cyst.

MATERIALS AND METHODS

Among the patients who had intracranial cystic lesions, 24 cases were diagnosed as hydatid cysts and underwent surgery between 1990-1998. Chest X-ray, abdominal ultrasound sonography and serologic confirmative tests were done in all cases in order to detect any systemic hydatidosis and to justify the diagnosis. Patients in whom cysts were ruptured during surgery and who had an additional cyst in any organ other than brain received mebendazole (800 mg daily for at least three months). A detailed recording was made of the presenting features, radiological findings, age, gender, treatment outcome, and surgery or postoperative complications.

RESULTS

Male to female ratio was 10/14 (41.7 and 58.3%), with age range 4 to 64 years (mean 24.4). Twenty patients (83.3%) came from rural areas, while four cases (16.3%) were from urban areas.

Duration of symptoms varied from 3 to 36 months, among which headache and vomiting were the most common symptoms, followed in order of frequency by papilledema, focal neurological deficits, seizure and speech disorder. Weakness, seizure and speech disorder occurred in 18 (85%), 6 (25%) and 4 (16.6%) patients, respectively. Table 1 summarized the different modes of presentation in our cases. Despite the large size of the cyst and considerable mass effect the patients remain in remarkable good condition. The locations of the cysts are shown in table 2.

Table 1. Clinical signs and symptoms in 24 patients with intracranial hydatid cysts

Presenting symptoms	Number of cases	Percentage
Headache	24	100
Vomiting	24	100
Papilledema	20	83
Weakness	18	85
Seizures	6	25
Speech disorder	4	16.6
Visual disturbances	4	16.6
Vault bulging	2	3.3

Table 2. Anatomic distribution of intracranial hydatid cysts in 24 patients

Location of the cysts	Number of cases	Cerebral hemispheres		Percent
		R	L	
Frontoparietal	7	4	3	28
Parietal lobe	4	1	3	16
Frontal lobe	3	1	2	12
Parieto-occipital	3	2	0	12
Frontotemporal	2	1	1	8
Parietotemporal	2	1	1	8
Occipital lobe	2	1	1	8
Temporal lobe	1	0	1	4
Thalamus	1	0	1	4
Total	25	11	14	100

Abbreviations: R, right; L, left.

Only one case had concurrent lung involvement. Lesions were almost always located in the cerebral hemispheres, of which 60% were on the right side. All but one of the patients had a solitary hydatid cyst; in the latter, two hydatid cysts were detected (one in the left thalamus and the second in the left parietal lobe). In 21 or 85.7% of cases, cysts were removed intact, in the remaining three patients cysts were ruptured during surgery, whom received mebendazole. There was no recurrence in the latter cases except for one with two cysts. Two patients died (one due to herniation and the second one with unknown cause) after surgery. Surgical mortality and postoperative complications occurred in 8.3 and 20.8%, respectively. Superinfection developed in 2 patients following the surgical intervention. Subdural hematoma and hydrocephalus each was detected in only one patient in postoperative computed tomography (CT).

DISCUSSION

Among the echinococcus species, the two most commonly associated with human disease are *E. granulosus* whose cyst has limiting membrane and *E. multilocularis* (alveolar) which is less common but more serious and is almost always fatal (3, 12). The latter lacks limiting membrane thus can grow aggressively.

Hydatid disease is prevalent in our country, especially in sheep grazing areas as East Azarbayjan

and Kordestan. Infestation by hydatid disease in human most commonly occurs in the liver (55-70%). The lung is the next most frequent site of cyst location (18-30%), followed by the kidneys, muscles, spleen, soft tissues, brain and bone (12,13). Liver and lung can be affected simultaneously in about 5-13% of cases (14).

Hydatid cysts constitute 3-4% of all intracranial space occupying lesions (15-20). Though the mortality directly due to echinococcosis is low (4-5%) but it can be a serious problem (21, 22). A male preponderance is reported only in adults (19, 22). In our study, though 65% of cases were children, in contrast to previous reports there was a female preponderance.

In cerebral hydatid cyst, there are two different histogenetic types, primary and secondary (26). Primary hydatid infection caused by embryos passing hepatic and pulmonary barriers. Primary intracranial cysts are the most common types and are commonly solitary. Secondary cysts are usually multiple which may follow embolization of ruptured cardiac cyst or spontaneous, traumatic and surgical rupture of a primary cyst in other organs. For this reason, in such cases the heart and other organs must be carefully investigated.

In adult, focal neurological signs like hemiparesis, speech disorder or seizures are usually the first to appear whereas in children the clinical picture is primarily raised intracranial pressure. Table 1 summarized the different modes of presentation in our cases. Headache and vomiting due to increased intracranial pressure were the most common presenting symptoms followed in order by neurological deficit and seizures.

Although hydatid cyst can occur anywhere in the brain, most of intracranial hydatid cysts are supratentorial in a cortical location in distribution of the middle cerebral artery (7, 27) and the parietal lobe is the commonest site as occurred in present series. Infratentorial hydatid cysts have been infrequently reported. The other less common reported sites are pons, cerebellum, basal ganglia, extradural, skull, cavernous sinus, ventricles and eyeball (28, 29). The locations of the cysts are shown in table 2.

In our series only one patient had concurrent lung

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involvement. Importantly, up to 20% of confirmed diseases have been associated with negative serology. False negative are also more frequent in disease involving the CNS and/or eyes (30, 31).

The diagnosis is most often reached when radiological studies show cystic space occupying lesions of the brain. In the present study most valuable diagnostic procedure was CT or magnetic resonance imaging (MRI) as reported in other series (32, 33). It was extremely difficult to diagnose hydatid cyst preoperatively until these methods became routine. Serologic testing is more specific, but less sensitive than most imaging modalities (1, 25). ELISA has up to 84 percent sensitivity (1). CT and MR imaging show a well defined smooth, thin walled spherical homogenous cystic mass with no edema and rim enhancement. Absorption values similar to that of CSF is the characteristic appearance on CT. MRI provides comprehensive information for accurate diagnosis. The cysts are hyperintense compared to CSF on PD (proton density) and isointense with CSF on T2 and T1-weighted images (34). Enhancement or edema is due to superinfection, rupture or leakages. MR scan may show a low signal intensity cyst wall and relation with surrounding structures is better delineated than on CT scan. We could not identify scolices on MR scan. Other reports on MR findings showed similar findings (30). Significant distortion of the brain parenchyma, ventricular displacement or hydrocephalus are common.

The radiological differential diagnosis is from other cystic lesions such as cystic astrocytomas, cerebral abscesses or arachnoidal cysts. Cystic astrocytomas show higher attenuation values than hydatid or arachnoidal cysts, due to higher protein content. The presence of surrounding edema and homogeneous enhancing solid component (mural nodule) of the tumor distinguishes it from a hydatid cyst. Abscesses show perifocal edema and peripheral enhancement. Rarely the cysts may show rim enhancement and enhancing nodule simulating a cystic astrocytoma or calcification in the wall of a degenerated cyst.

In summary, Iran is one of the countries where echinococcosis is a common parasitic disease especially in sheep and cattle raising areas. Primary

hydatid cyst of the brain especially in adults is rare and can pose various diagnostic problems. Hydatid cyst should be included in the differential diagnosis when a cystic brain lesion is found in patient from an endemic echinococcosis area. Extensive search for parasites in some other organs is indicated if a hydatid cyst is found in the intracranial cavity. CT scanning is extremely useful for diagnosis, but MRI visualized cyst location better than CT.

Clinical signs and symptoms are related to the site and size of the cyst. Treatment of choice is surgical with complete removal of the cysts without spillage of their contents whenever possible. Most common complication is a rupture of the cyst into the subarachnoid space, which leads to wide spread dissemination followed by severe anaphylactic response. In this study cysts were removed intact in 21 (%) of 24 patients, in the remaining 3 patients, however, the cysts were ruptured during surgery. In the present series 4 patients received medical treatment (mebendazole), three of them had cyst rupture during surgery and one had liver hydatid cyst. There was no recurrence in these 3 patients except for one with two cysts. Two patients died after surgery. The cause of death in one patient was upward herniation, in the next patient the cause was not available.

Preoperative diagnosis of hydatid disease is essential, because the rupture may result in an anaphylactic reaction with circulatory collapse and cardiac arrest, or dissemination of the cyst may result in recurrence. Removal of giant cysts, which have a thick wall, is easier than smaller ones, which are prone to rupture. Intact delivery of deeper ones is a difficult problem. In the present series 3 cases experienced rupture: one located within the left thalamus, the other two had deep sylvian fissure (fronto-parietal) and deep parietal location. Superinfection developed in 2 patients following surgical intervention. Subdural hematoma was observed on the postoperative CT scans of one patient.

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