Case report

A rare case of primary multiple hydatid cysts of the brain in a 10-year-old child

M.M.K Boujan[1] and Z.S.A. Jalal[1]

Introduction

Echinococcus is a cestode. The most common *Echinococcus* species causing human infection are *E. granulosus* and *E. multilocularis* [1].

The commonest sites for cysts are the liver (59%–75%), followed in frequency by lung (27%), kidney (3%), bone (1%–4%) and brain (1%–2%). Other sites such as the heart, spleen, pancreas and muscles are very rarely affected [2]. Multi-organ involvement without liver cysts is reported [3].

Brain cysts caused by *E. granulosus* are rare; they occur during childhood in endemic areas [4]. Cerebral hydatid cysts are usually supratentorial and tend to occur in the middle cerebral artery distribution [5]. The cyst's predilection for white matter is undisputed, but the exact reason for this is unknown; some believe it is because of the arterial distribution [6].

Cerebral hydatid cysts are solitary. Multiple hydatid cysts in brain parenchyma are very rare and result from spontaneous, traumatic or post-surgical rupture of cysts in the brain or elsewhere; with embolization of hydatid to brain, multiple cysts may arise in the brain itself [5–8].

Cerebral hydatid cysts are classified as primary or secondary. Primary intracranial hydatid cyst results from direct infestation of the brain by the larva without involvement of other organs such as liver and lungs. Primary cysts are fertile, containing brood capsules and scolices, and their rupture may result in recurrent intracranial hydatid cyst. Primary cysts are usually solitary. Primary multiple cerebral hydatid cysts are quite unusual, resulting from multiple larvae infecting the brain after bypassing the liver and lungs [8,9].

Secondary cysts are generally multiple and result from the rupture of hydatid cysts in the brain or in other organs of body. These cysts lack germinal membranes and are infertile, with negligible risk of recurrence after rupture [6,7,10]. Here we report a rare case of primary multiple hydatid cysts of the brain in a 10-year-old child, with a total of 73 cysts removed from his cerebrum and 5-year follow-up. Written informed consent was obtained from the patient's parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by the *Eastern Mediterranean Health Journal's* Editor-In-Chief.

Case report

A 10-year-old boy was admitted to paediatric hospital with a history of single attack of fit, no previous history of epilepsy, no history of jaundice, other liver disease or lung disease. His parents gave a history of unexplained fever for several days.

The patient was referred to the neurosurgery department. Neurological examination revealed bilateral papillodema, lower face weakness but otherwise normal motor power. CT of the brain showed a left parietal–temporal cystic lesion, minimally enhanced after contrast. MRI showed multiple cysts in a larger cyst.

The patient underwent a craniotomy, and 67 cysts were taken out; there were no more visible cysts in the brain. The part of cerebral cortex over the cyst hung loosely into the resulting cavity.

Postoperatively the child was right-sided hemiplegic of grade zero power of lower face, upper and lower limbs, without loss of cortical sensation.

Hydatid disease was confirmed histopathologically.

Within three months the patient gradually improved to full power except for power grade three in the right hand finger movements and right foot dorsiflexion.

Patient was put on an albendazole regime, 200 mg tablet twice daily of 28 days on, 14 days off medication, for three cycles. Carbamazepine 200 mg was given once daily.

After two years the patient, who had been seizure-free for this period, presented with another attack of tonic–clonic fit. A CT brain scan revealed recurrent hydatid cysts.

A craniotomy was done, and six cysts were evacuated. Albendazole cycles were restarted with carbamazepine 200 mg twice daily.

Eight months later, the patient had a clear follow-up CT brain scan.

[1] *University of Sulaimani School of Medicine, Sulaimani, Iraq (Correspondence to M.M.K Boujan: mazinboujan@yahoo.com).*
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The patient had a third seizure 16 months after his last CT, 24 months after his second craniotomy. He was admitted to hospital; his CT brain scan was negative for recurrence. The antiepileptic dose was increased to 200 mg three times daily.

Currently the patient is well, on carbamazepine 200 mg three times daily and on follow-up for another two years of antiepileptics.

During the course of his illness the patient's multiple abdominal ultrasounds, chest X-ray, and cardiac echo were normal.

Discussion

Cerebral hydatid disease is usually in the form of one single cyst that might reach a large size. Multiple cysts rarely occur, caused by accidental spillage during surgery or trauma or even spontaneously; these are so called multiple secondary cerebral hydatid cysts of the brain. Primary multiple cerebral hydatid cysts of the brain are extremely rare as multiple larvae directly invade the brain [8].

Apart from the common knowledge that hydatid disease is endemic in Iraq [11], no reason was found in this particular patient for the means of infection, as the patient was living in an urban area, and there was no history of contact with the primary host, dogs. However, picnics in rural areas of Kurdistan are popular.

E. granulosus (cystic echinococcosis) is endemic in Mediterranean Europe, the Middle East, North Africa, Australia and South America [12]. *E. multilocularis* (alveolar echinococcosis) is endemic in Alaska, central Europe, Turkey and China [1].

Human cystic echinococcosis (hydatidosis) is a disease caused by the larval stages of the dog tapeworm *E. granulosus* and it is principally maintained in a dog–sheep–dog cycle [13].

Intracranial hydatid cyst is rare even in countries where the cestode is considered endemic. Hydatid cyst of the brain accounts for 1%–2% of all intracranial space occupying lesions; 50%– 75% of intracranial hydatid cysts are seen in children [14,15].

In the countries of the Eastern Mediterranean Region, echinococcosis is one of the major zoonotic parasitic diseases from Morocco to Egypt. Both cystic and alveolar echinococcosis have been reported from these areas. However, cystic echinococcosis is more common and has been reported from all countries in the Middle East and Arabic north Africa. Alveolar echinococcosis is less prevalent and has been reported only from Islamic Republic of Iran, Turkey, Iraq and Tunisia. *E. granulosus* is highly prevalent in Islamic Republic of Iran, Turkey, Iraq, Morocco, Tunisia and Libya. In Oman, it is endemic with low prevalence. Cystic echinococcosis has a very low level in Cyprus. Hydatid cyst is commonly found in sheep, cattle, goats and camels throughout the areas of Middle East and Arabic north Africa. Sheep are infected mostly in these regions. The disease is hyperendemic in Islamic Republic of Iran, Turkey, Iraq, Jordan, Morocco, Libya, Tunisia and Algeria, and endemic in Egypt. Studies on the strain specificities of *E. granulosus* revealed that sheep strain is present in sheep, goats, cattle, camels and humans, and the camel strain in camels, sheep and cattle as well as humans. Dog–sheep strain seems to be more prevalent in Islamic Republic of Iran and Jordan. However, horse strain has been reported from Jordan. In Egypt the human cases reported are of camel–dog strain [16].

In Iraq, studies showed that housewives, labourers and farmers appear to be at the greatest risk of infection [17]. In another study the disease was found more prevalent in active workers age 20–50 years, with higher incidence in women [18]. In countries where infection is endemic, females and children are affected more, probably due to more frequent contact with infected animals [19].

The disease is endemic in the Syrian Arab Republic. The north of the country is the main area for raising livestock and of nomadic tribal life [6].

Cyprus had a control programme against *Echinococcus granulosus* that started in 1971–74. Full control was declared in 1985, but subsequent studies in 1993–96 revealed that 20% villages were infected so the control programme was reintroduced [20]. Unilocular hydatid disease, *E. granulosus*, is also endemic in Sardinia. In this region there are foci of human infection, notably in the sheep-rearing areas [21].

In Australia, hydatid disease has historically been sustained in a farm-dog to sheep cycle. When dogs are fed the offal of infected sheep, they become infected with *E. granulosus*. The disease is most common in the sheep-farming areas of New South Wales, the Australian Capital Territory, Victoria, southwest Western Australia and eastern Queensland. It is believed to occur in South Australia but its status is unknown. It has also been found in cattle populations in the Kimberley region of Western Australia, in northern Queensland and near Darwin in Northern Territory. Scientists believe that *E. granulosus* arrived in Australia soon after European settlement, probably in infected sheep or dogs. Its distribution is determined largely by climate [22].

In South America, Cystic echinococcosis constitutes an important public health problem in Peru [23]. The economic impact of the disease has been assessed in South American countries where it is endemic. A recent study in Argentina, Brazil, Chile and Uruguay demonstrated major human and animal losses in all four countries [24].

According to WHO, the hepatic cysts (as well as the cerebral cysts) are classified by their MRI appearance as one of three fertile types as follows: fertile active cysts that appear as unilocular cysts with no visible wall or a clear visible wall (CE1), a unilocular mother cyst with multiple vesicles arranged peripherally along the cyst wall (CE2) and the transitional form (CE3) showing multiple daughter cysts entirely filling the maternal cyst. There are two types of inactive cyst that have lost their fertility: a "ball of wool" with collapsed membrane or detached membrane with water-lily sign (CE4) and calcified lesions (CE5) [10].

Our patient presented with the least frequent presentation in literature: seizure. Symptoms and signs of intracranial hydatid disease are headache and vomiting in 91%, visual field defect in 35%, papillodema in 74%, hemiparesis in 62%, mental change in 22% and seizure in 20% [6,25].

In our first craniotomy, we were meticulous not to rupture any cyst while operating. Despite this and the fact that we used albendazole postoperatively, the patient had a recurrence, and a second craniotomy was needed with readministration of albendazole, which finally paid off with clear subsequent follow-up CTs.

The most effective treatment of hydatid disease is surgery, followed by a course of medication to eradicate any remaining tapeworm larvae that might be in the body [22,26]. Albendazole was found to have favourable effects in patients with severe, inoperable hydatid disease in one report [27]. Two other reports stated that following albendazole administration a clear reduction in the size and number of hydatid cysts in the brain was noticed [28,29]. Recurrence of hydatid disease after surgery and postoperative albendazole is reported in hepatic hydatid disease [30].

Recumbent antigen is now used as a vaccine for sheep against echinococcosis that has shown up to 90% protection; however, the use of the vaccine does not mean that hydatid disease will be eradicated as vaccinating livestock will not reduce transmission of hydatid disease in the wild. The vaccine probably will have its biggest impact in those parts of the world where hydatid disease is most prevalent, the countries mentioned above. On the other hand, such a success encourages investigating its use in humans [22,31].

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