The Gigant primary cerebral hydatid cyst with no marked manifestation: a case report and review of literature

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Abstract. – Hydatid cyst is a parasitic infestation that rarely affects the brain. A 14 year-old child with a large mass of intracranial hydatid cysts (122x110x98 mm), but no symptoms except for an intermittent headache for 2 months is presented. Triple cysts were removed after suitable craniotomy. The literature was reviewed and the case was compared with the published reports.

Key Words: Brain, Children, Hydatid cyst.

Introduction

Hydatid cyst is a parasitic infestation caused by the larval stage of the different species of the tapeworm Echinococcus genus. The most common form worldwide including Turkey is cystic echinococcosis that is chiefly caused by Echinococcus granulosus. It is endemic in Mediterranean countries, the Middle East, South America, and Australia.

Like many other parasitic infestations, the course of Echinococcus is complex. The worm has a life cycle that requires definitive hosts and intermediate hosts. Definitive hosts are normally carnivores such as dogs and cats while intermediate hosts are usually herbivores such as sheep or cattle. Echinococci are transmitted to intermediate hosts via the ingestion of eggs as humans do. Whereas, they are transmitted to definitive hosts by means of eating infected, cyst-containing organs. Humans are accidental intermediate hosts that become infected by handling soil or dog excrement that contains eggs or by ingestion of food contaminated by the ova of the parasite.

The most commonly affected organs are the liver and lungs; the hydatid cyst of the central nervous system is unusual with a reported frequency of 1-2% of all cases with hydatid cyst disease.

The aim of this study is to emphasize that any lesion, even though it has reached a massive size, may not be able to show neurological symptoms. To the best of our knowledge, this is the largest primary cerebral hydatid cyst with no symptoms to come across in the English literature.

Case Report

A girl of 14 year-old presents who is complaining of a blunt headache in the frontal region. While investigating the etiology the headache, her cranial CT revealed a huge intracranial mass that extraordinarily gives no more symptoms than a headache for a couple of months with no need for any medication. She was in a good general state of health, fully conscious and oriented, and her neurological examination did not reveal any neurological deficit. Although the lesion embedded within the dominant hemisphere, she was intact in speech and did not have any symptoms of increased intracranial pressure such as nausea or vomiting. Only a minor degree of papilledema was shown by fundoscopic examination on the lesion side. Magnetic resonance imaging of the skull with and without contrast showed well-defined, very large sized, extra axial cysts with no enhancement, extending from the left frontal region to the occipital (Figures 1, 2). History of direct contact with dogs is also not available.

After written her consent letter, she was operated on by a left fronto-temporo-parietal craniotomy. The cyst was removed by the Dowling-Orlando technique with the aid of gravity without rupture (Figure 3). Histopathological examination confirmed as the hydatid cyst and medical therapy was completed by albendazole for a couple of months under serological controls.

Discussion

Hydatid cyst disease is endemic in the Middle East and Mediterranean countries in addition to South America, North Africa and Australia. Intracranial involvement is rare, but more commonly
seen in pediatric population, and 80% of patients in the recent series were children. This high incidence in children is probably related to patent ductus arteriosus. As is the case, a history of direct contact with dogs is not available in all the cases, because the infection can usually be acquired by ingesting whatever material was contaminated.

Intracranial cysts are more frequently settled in the supratentorial compartment and the parietal lobe is the most preferable site. All four cases reported by Dharker et al and three out of five cases of intracerebral hydatid cysts reported by Balasubramaniam et al had parietal lobe involvement. The other less common sites reported are skull, cavernous sinus, eyeball, pons, skull, extradural, cerebellum and ventricles. In current case, the multiple lobes involvement is seen due to massive enlargement (Figures 1, 2).

Intracranial hydatid cysts are commonly solitary. Multiple cysts are rare. Onal et al reported only three multiple cysts in their series of 33 cases and Lunardi et al found 2 cases in their series of 12 cases. Present case was also multiple with triple large cysts.

Intracranial hydatid cysts may also be classified as primary or secondary. The primary cysts are formed as a result of direct infestation of the larvae in the brain without demonstrable involvement of other organs. In primary multiple cysts, each cyst has a separate pericyst with brood capsule scolices and these originate from multiple larvae affecting brain after crossing the gastrointestinal tract, liver, lungs and right side of heart without affecting them. The primary cysts are fertile as they contain scolices and brood capsules, hence rupture of primary cyst can result in recurrence. The secondary multiple cysts results from spontaneous, traumatic or surgical rupture of the primary intracranial hydatid cyst and they lack brood capsule and scolices. The secondary intracranial hydatid cysts are, therefore, infertile and the risk of recurrence after their rupture is
negligible. Primary multiple cysts as in the case are uncommon and isolated case reports of primary multiple hydatid cysts have appeared in the literature. Nurchi et al. found only 11 reported cases of primary multiple hydatid cysts while reviewing the literature.

The patients with intracranial hydatid cysts usually present with focal neurological deficit and features of raised intracranial pressure; the latter may be due to the large size or due to interference with pathways of cerebrospinal fluid flow. Ersahin et al. observed that 18 out of 19 cases presented with raised intracranial pressure and four of those cases had seizures. In a other study, all patients had focal neurological deficits and features of raised intracranial pressure and two patients had seizures. The current case was characterized by no neurological symptoms even though the cyst was in huge dimensions over eloquent areas.

For simple cases of cystic echinococcosis, the most common form of treatment modality is surgical removal of the cysts combined with chemotherapy using antihelmintic before and after surgery. However, if there are cysts in multiple organs or tissues, or the cysts are in risky locations, surgery may become impractical. The aim of surgery is to excise the cyst in entirety without rupture to prevent recurrence and anaphylactic reaction. Various surgical options as summarized by Arana-Iniquez include: puncture and aspiration of the cyst fluid through a small hole in the cyst wall, cortical incision over cyst and expulsion of hydatid cyst by insufflations of air in the contralateral ventricle, and the most commonly performed procedure, to give birth to the intact cyst by irrigating saline between the cyst wall-brain interface. This can be possible thanks to minimal adhesions around the cyst wall. The case was operated on with the aid of gravity by the means of the operating table by the last procedure.

Only a few reports are available mentioning the efficacy of drug therapy. Golematitis et al. analyzed 44 patients who were treated with albendazole and found that the large cysts decreased in size, while the smaller ones disappeared. Ersahin et al. reported better effectiveness of the drug therapy in recurrent cases and in cases with rupture at the surgery. In current case, the medical treatment was applied just after surgical removal in accordance with the general approach.

Common magnetic resonance imaging (MRI) findings of hydatid cyst are well-defined, smooth thin-walled, spherical, homogeneous cystic lesions with no contrast enhancement, no calcification, and no surrounding edema (Figures 1, 2, 3). A tricky question is why such a large lesion is not presenting any major symptoms in spite of occupying the eloquent areas in the dominant hemisphere. This is only possible with a gradually growing and slowly overwhelming mass, which lets neural tissue function.

Conflict of Interest
The Authors declare that there are no conflicts of interest.

References