Case Report

Multiple intracranial hydatid cysts in posterior fossa in an adult—A case report

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Abstract

Hydatid cyst is an uncommon parasitic disease caused by larval stages of Echinococcus granulosus. The liver is the most frequently affected organ followed by the lungs and the spleen. Intracranial hydatid cysts are uncommon and occur mostly in supratentorial region. It can present with nonspecific symptoms and can be difficult to diagnose, thus regardless of unusual clinical presentation and unusual location of cystic lesion in brain, it is crucial to keep hydatid cyst as one of the differentials. We describe a case of a 28-year-old male who presented with headache, vomiting and cerebellar signs. MRI showed multiple cystic lesions in posterior fossa with asymmetrically dilated posterior horn of left lateral ventricle. Biopsy from one of the cystic lesions from posterior fossa was performed which confirmed the diagnosis of hydatid cyst. Patient was started on Albendazole and subsequently planned for surgery.

Introduction

Hydatid cyst is an uncommon parasitic disease caused by larval stages of Echinococcus granulosus. Humans are the intermediate host that contract the disease by consuming contaminated embryonated eggs of Echinococcus. These eggs produce oncosphere in the intestine which penetrates the walls of intestine, migrates into various organs via systemic circulation and develop into cyst. The liver is the most frequently affected organ followed by the lungs and the spleen. However, the central nervous system is seldom affected [1]. Intracranial hydatid cyst accounts for approximately 1-2% of all cases [2]. Children and young adults are more likely to have intracranial hydatid diseases than adults. Most intracranial hydatid cyst are unilocular and solitary and they most frequently develop in the cerebral hemispheres, especially in the region of the middle cerebral artery [3].
In this case report, we describe a unique instance of multiple intracranial hydatid cysts located in posterior fossa in a 28-year-old male who exhibited nonspecific clinical findings which was confirmed by findings in imaging and histopathology.

**Case report**

A 28-year-old male patient presented to our hospital with one year history of intermittent headache which had gotten worse over previous one week and was accompanied by nausea, vomiting and diplopia. Additionally, he also reported gait disturbances during the previous 4 months. Neurological examination revealed cerebellar ataxia with normal sensations, motor functions and reflexes. Physical examination revealed approximately 4.5 × 3 cm sized swelling over occipital region on left side which was compressible, fluctuant and non-tender. Routine blood analysis, chest radiograph and abdominal ultrasonography findings were normal. Considering the patient’s signs and symptoms, MRI was ordered which showed multiple small cystic lesions along with a large cystic lesion in posterior fossa with mass effect in adjacent cerebellum (Figs. 1 and 2). A differential diagnosis of cystic lymphangioma was considered.

Since MRI findings were not conclusive for a definitive diagnosis biopsy was planned. Biopsy sample was obtained from the largest described cystic component in the posterior fossa which revealed that the cystic structure was lined by an outer layer of avascular eosinophilic laminated chitinous layer with an innermost germinal layer (Fig. 3A). Few inflammatory cells and loosely arranged fibro-collagenous tissues were also noted (Fig. 3B). These findings in histopathology were compatible with hydatid cyst.

CT scan of chest and abdomen was then ordered to search for evidence of hydatid cyst elsewhere which revealed no abnormal findings. With the diagnosis of intracranial hydatid
Dense cystic lesions in posterior fossa demonstrated complete suppression, while large lobulated component not showing suppression. (B) Postcontrast axial images show no enhancement of the lesions. (C & D) DWI/ADC sequences do not show areas of diffusion restriction within the lesions.

cysts, the patient was started on oral Albendazole and was subsequently planned for surgery.

**Discussion**

Central nervous system involvement of hydatid cyst is seldom encountered accounting for 1-2% of the incidence worldwide [2]. The condition is mostly encountered in children and young adults with radiological presentation of solitary unilocular cyst in supratentorial region [3].

The causative organism for hydatid cyst is a parasite called Echinococcus granulosus. The adult Echinococcus granulosus lives in the definitive host’s small intestine. Gravid proglottids release eggs that are infectious and are passed in the feces. After being consumed by a suitable intermediate host, eggs hatch in the small intestine, releasing 6-hooked oncospheres that penetrate the intestinal wall and migrate through the circulatory system into various organs, particularly the liver and lungs. The oncosphere develops in these organs into a thick-walled hydatid cyst that gradually enlarges, producing protoscolices and daughter cysts. The definitive host becomes infected by ingesting the cyst-containing organs of the infected intermediate host. Following ingestion, the protoscolices evaginate, attach to the intestinal mucosa and proliferate. Humans are unnatural intermediate hosts who become infected after consuming eggs. Oncospheres are released in the intestine, and hydatid cysts form in a variety of organs. If cysts rupture, the liberated protoscolices may form secondary cysts elsewhere in the body (secondary echinococcosis) [4].

After passing through the liver and lungs hydatid cyst rarely reaches the intracranial cavity. Intracranial hydatid cysts are frequently located in supratentorial compartment, usually temporo-parieto-occipital, in the distribution of the
terminal branches of the middle cerebral artery [2]. In our case, there were multiple cystic lesions in posterior fossa with most of the lesions showing complete suppression on axial FLAIR image. In addition, there was a lobulated cystic lesion of T1 iso signal intensity with low signal intensity internal content which was subsequently biopsied and revealed findings suggestive of hydatid cyst.

The treatment of hydatid cyst is exclusively surgical. Surgeon must be patient enough to rely on hydrostatic pressure of saline flushed into the cavity. Lowering the head end of the patient also facilitates the delivery of the cyst by gravity [5].

The first description of the technique of hydrodissection method was made in 1929 by Dowling and Orlando. It entails the following 5 steps: skin incision in large flaps; craniotomy; cortisectomy of at least three-fourths of the cyst diameter; application of warm 3% hypertonic saline to the surgical borders separating the brain from the cyst; and lastly, delivery of the cyst. The cortisectomy and the determination of the plane between the cyst wall and cortex are the two most crucial steps in the surgery.

It is regarded as a safe method for complete cyst removal, as demonstrated in our case, avoiding intraoperative rupture of the cyst, spillage of cyst contents, anaphylactic shock, and immediate mortality, as well as disease recurrence later on [6].

Albendazole can be given for a variety of reasons including sterilization of the cyst, reduction of the risk of anaphylaxis, reduction of tension in the cyst wall and reduction of the rate of recurrence [7].

**Conclusion**

Hydatid cysts are rarely encountered in intracranial cavity and posterior fossa location is even rarer. A cystic lesion in intracranial cavity should alert the clinician to consider hydatid disease as one of the possible diagnosis which will help in prompt and appropriate management of the condition.

**Patient consent**

Consent from the patient was taken on written form for case report and using the MRI images in any journal after explaining on his own language.

**References**

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