CASE REPORT



Giant hydatid cyst in the posterior fossa of a child

Fakhr Fakhouri, Abdelwahed Ghajar¹, Nihad Mahli², Nihad Shoumal³

Division of Neurosurgery, Department of Surgery, ¹Associated Professor of Neurosurgery and Chief of Division of Neurosurgery, Department of Surgery, ²Professor of General Surgery and Chief of Department of Surgery, ³Neurosurgery resident, Division of Neurosurgery, Department of Surgery, Aleppo University Hospital, Aleppo, Syria

ABSTRACT

The hydatid cyst (HC) is endemic in Mediterranean region including Syria. The central nervous system is involved in 2-3% of cases. HC in cerebellum is very rare. We report a case that presented as an emergency for high intracranial pressure and deteriorating Glascow coma scale. Close monitoring and precise surgical management using Dowling's technique resulted in very good outcome with full recovery. We highlight the need for very careful surgical treatment because cyst rupture and secondary hydatidosis due to spillage of the cyst contents can dramatically worsen the outcome. HC should be taken into consideration in countries where hydatid infestation is endemic.

Key words: Children, Dowling's technique, elevated intracranial pressure, hydatid cyst, posterior fossa

Introduction

Hydatid infestation is endemic in many parts of the world including the Mediterranean region, Middle East, Southern Asia, Latin America, and Australia. [1-3] However, it is now also seen in western countries because of travel and migration. [4] An intracranial hydatid cyst (HC) is a relatively rare entity, accounting for only 1–2% of all intracranial space-occupying lesions. [1,2,5] Cerebral HCs are most frequently supratentorial while those located in the posterior fossa are very rare. [6,7] Here, we report a case of a child presented with acute hydrocephalus due to a large posterior fossa HC. Close monitoring and proper surgical technique resulted in very good outcome.

Case Reports

A 5-year-old girl presented with a 3-month history of progressive headache, intermittent vomiting, and difficulty in walking. The headache was generalized and worsened in the morning. On admission, she was showed a deterioration in the level of consciousness (her Glascow coma scale was 12) with bilateral admitted for high intracranial pressure with Glascow coma

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Address for correspondence:

Dr. Fakhr Fakhouri, Department of Neurosurgical, Aleppo University Hospital, Aleppo, Syria.

E-mail: fakhrf@msn.com

scale being 12. She had bilateral papilledema. Brain computed tomography (CT) revealed a right sided large cerebellar cystic mass surrounded by mild edema. CT also showed marked ventriculomegaly and periventricular edema [Figure 1]. The patient was admitted in the Intensive Care Unit and started on steroid. About 24 h later, she became fully alert and was examined thoroughly. She had a wide gait with a tendency to fall to the right side. Positive cerebellar signs (finger nose sign, heel knee sign) were confined to the right side. Routine blood analysis was within normal limits. Abdominal ultrasonography revealed two small cysts in right hepatic lobe measuring 9 and 10 mm in diameter [Figure 2]. Chest X-ray was normal [Figure 3]. Brain magnetic resonance imaging (MRI) showed a cystic lesion in the posterior fossa that was hypointense on T1-weighted sequences with minimally enhancing cyst wall [Figure 4]. On T2-weighted sequences, it was hyperintense [Figure 5]. MRI also confirmed pericystic edema better seen on flair sequences [Figure 6].

The day after, she underwent suboccipital craniectomy, and a relatively large cerebellar HC (approximately 6 cm in diameter) was removed using hydrodissection as described in Dowling's technique [Figure 7]. The diagnosis was confirmed by pathological examination of the cyst. The postoperative course was uneventful, and the patient was discharged 10 days later with complete neurological recovery. Albendazole was prescribed for 4 weeks.

Discussion

Hydatid disease is caused by encysted larvae of the dog tapeworm *Echinococcus granulosa*. It is endemic in Middle East, Mediterranean countries, South America, North Africa, and Australia especially in a rural area. Man is infected either by eating contaminated food or by direct contact with an infected dog. Central nervous system involvement occurs in only about

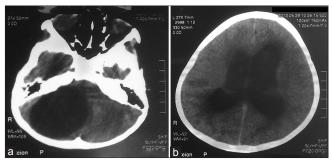


Figure 1: Computed tomography scan showing (a) posterior fossa cyst (left), (b) marked ventriculomegaly and periventricular extravasation (right)



Figure 3: Chest X-ray was normal

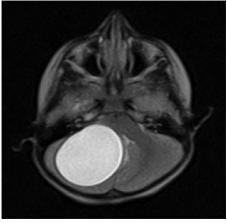


Figure 5: T2-weighted magnetic resonance imaging showing hyperintense cystic component

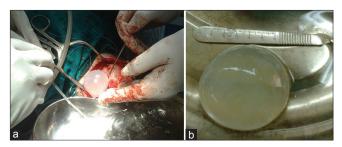


Figure 7: Intra-operative view showing the cyst (a) as it is delivered (left), and (b) when compared to ruler scalpel (right)

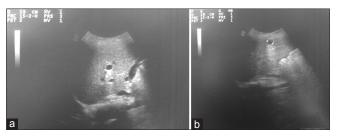


Figure 2: (a and b) Abdominal ultrasonography showing two small hepatic cysts

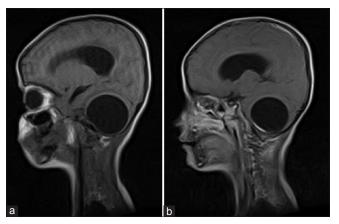


Figure 4: T1-weighted magnetic resonance imaging (a) before (left) and (b) after (right) contrast administration

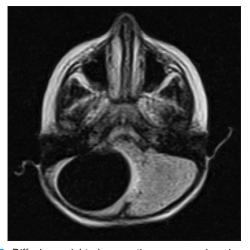


Figure 6: Diffusion-weighted magnetic resonance imaging showing mild edema around the cyst

3% and produces cerebral cysts that are usually confined to the white matter. [8] Cerebral HC is more common in the pediatric population, probably related to a patent ductus arteriosus. [2,9] Intracranial HC is most commonly located in the supratentorial compartment, [10] and less commonly in the cavernous sinus, [11] eyeball, [12] pons, [13] and cerebellum. [14]

Cerebral HC's are slowly growing so it is frequently diagnosed several months or even years after the onset of symptoms, which are usually attributed to high intracranial pressure such as headaches and vomiting, or less commonly to focal neurological deficit.^[15] Rarely could HC present as an emergency as in our case. The growth rate of HC has been reported between 1 and 5 cm/year to 10 cm.^[16] Solitary intracranial HC's are common than multiple intracranial cysts.^[17]

Computed tomography and MRI reveal distinctive features of solitary HC. HC generally does not enhance, although rim enhancement may occur if there is an inflammatory reaction. [8] In our case, there was little enhancement. However, large rounded cystic lesion which is isodense and isointense, respectively, to cerebrospinal fluid with slight to no rim enhancement should raise the suspicion of HC. Brain edema is usually minimal. Magnetic resonance (MR) spectroscopy and MR diffusion-weighted imaging might help in the diagnosis of intracranial HC in difficult cases. [18,19]

A variety of surgical techniques is used for removal of the HC's. [20] The most important complication of the surgical treatment is secondary hydatidosis due to spillage of the cyst contents. The popular technique is Dowling's technique of hydrodissection in which normal saline irrigation is used with mild force between the cyst wall and brain interface in order to deliver the cyst intact. [21] This is often possible because the adhesions around the cyst wall are minimal. In our case, we performed this procedure successfully. Adjunctive medical therapy with albendazole may be used. In our patient, it was necessary because of concomitant hepatic cyst. [22]

Conclusion

Even though HC in cerebellum is very rare, in an endemic area, it should be considered in the differential diagnosis of posterior fossa cysts. Dowling's technique is the safest surgical technique as cyst can be delivered intact with the best outcome.

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