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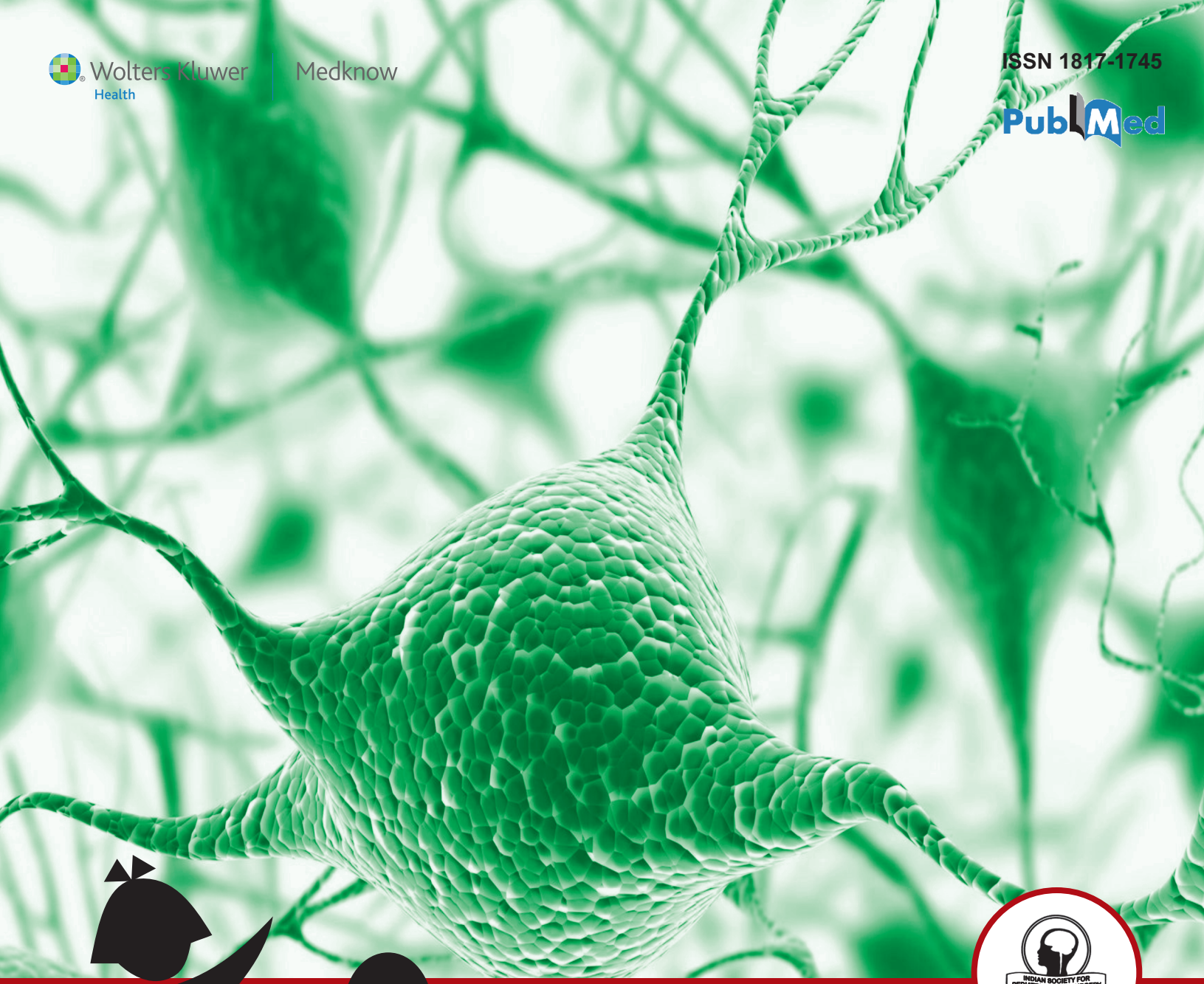

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Case Report

Intracranial hydatid cyst is a rare cause of midbrain herniation: A case report and literature review

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ABSTRACT

Hydatid disease is a parasitic infection affecting the brain in about 2% of the cases. Brain involvement is most commonly observed in children. Here, we report a 13-year-old male patient who presented with headache, nausea, and vomiting. Before cranial computed tomography (CT) was performed, the patient had generalized epileptic seizures. He was disoriented, and had anisocoria with dilatation of the right pupilla. CT showed a cystic lesion of 10-cm diameter in the right temporoparietal region that had caused a shift of the midline structures to the contralateral side; an urgent operation was performed as there were signs of midbrain herniation.

Key words: Hydatid cyst, mass effect, midbrain herniation, porencephalic cyst, urgent operation

Introduction

Human hydatid disease is caused by the larval form of *Echinococcus granulosus*. The definite host of *E. granulosus* is dogs. Intermediate hosts are typically sheep; however, the disease can occur less frequently in humans. A history of direct contact with dogs is not available in all reported cases, and infection can be acquired by the consumption of contaminated food and milk.[1,2] After ingestion of contaminated food, the embryos migrate through the portal system to the liver, and later the lungs.[3] The liver is the primary organ affected by the disease. Intracranial hydatid disease is rare, with a reported incidence of 1-2% of all cases with hydatid disease.[4,5] Cerebral hydatid cysts are more common in the pediatric population, with 80% of the affected patients being children; moreover, the cysts are most frequently observed in the supratentorial region and parietal lobe.[6] The high incidence in children is due to patent ductus arteriosus[7] or other valve dysfunctions.[8] Different/various growth rates of the cyst in the brain have been reported.[8-14] The main treatment is the complete surgical removal of the cyst without rupture, and the most commonly used surgical technique is the Dowling method modified by Arana-Iniguez.[2,3]

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Case Report

A 13-year-old male patient was admitted to the emergency department with headache, nausea, and vomiting. Neurologic examination revealed bilateral papilledema. Before cranial computed tomography (CT) was taken, the patient had generalized epileptic seizures. He was disoriented, and had anisocoria with dilatation of the right pupilla. His Glasgow Coma Scale value was 7 (E1 M5 V1). CT showed a solitary cystic lesion with a diameter of 10 cm, which was hypodense, sharply demarcated, without perifocal edema, and caused a shift of the midline structures to the contralateral side. The cyst was located in the right temporoparietal region [Figure 1].

Because of the midbrain herniation signs, urgent operative intervention via a large craniotomy was performed. Once the dura mater was opened, a linear corticotomy was made. After visualization of the cyst wall, 0.9% saline solution was used to dissect the hydatid cyst from the surrounding brain to facilitate cyst removal (Dowling method modified by Arana-Iniguez). Valsalva maneuver was also applied to

facilitate the removal of the cyst. The patient's head was lowered to further ease the cyst removal by gravity. The hydatid cyst was removed without rupture. The dura was closed in a watertight fashion after filling the remaining cavity with 0.9% saline solution [Figure 2].

Postoperative thorax and abdominal CT revealed no other foci. Fifteen days after the operation, a left hemiparesis occurred and cranial CT scan revealed a brain compressing subdural hygroma [Figure 3a]; therefore, we inserted a subdural-peritoneal shunt. Based on cranial CT performed during follow-up, the subdural-peritoneal shunt was effective and no complaints were reported [Figure 3b]. The neurological examination was normal. In the same CT examination done for follow-up, we noted that the site from where the hydatid cyst was removed was flask-shaped, relaxed space, and filled with cerebrospinal fluid (CSF) [Figure 3b]. However, in the follow-up at the fourth month after intracranial operation, we saw a distended cyst that had replaced the flask and relaxed CSF-filled space from where the hydatid cyst was removed; interestingly, we noted a jet-flow with flow void effect as an influx. This appearance suggested that a one-way valve mechanism had developed that filled the space [Figure 4].

In Figure 4, the jet-flow and flow void of the high-speed CSF can be easily observed. This setup demonstrates the one-way valve mechanism of CSF flow, which causes enlargement of the porencephalic cyst. The patient underwent a third surgical procedure, and fenestration of the cyst to the subdural space was performed. A postoperative follow-up cranial CT scan showed no signs of the porencephalic cyst [Figure 5]. Postoperative albendazole therapy was then applied for 4 months (10 mg/kg per day). The patient was discharged following normal neurological exams. Currently, the patient is healthy and attending school without any epileptic seizures.

Discussion

Human hydatid disease is caused by the larval form of *E. granulosus*. The liver is the primary organ affected by the disease. Intracranial hydatid disease is rare, with a reported incidence of 1-2%.[4,5] A history of direct contact with dogs is not available in all reported cases, and the infection can be acquired by consuming contaminated food and milk. A cerebral hydatid cyst is more common in the pediatric population, with 80% of the patients being children. This high incidence in children is primarily due to patent ductus arteriosus[7] or other valve dysfunctions.[8]

Intracranial hydatid cysts are more frequently located in the supratentorial region, with the parietal lobe being the most common site.[6] Hydatid cysts typically involve the middle cerebral artery territory, with other less common sites being the skull, cavernous sinus, eyeball, pons, cerebellum, ventricles, and interpeduncular cistern.[15] Cerebral hydatid

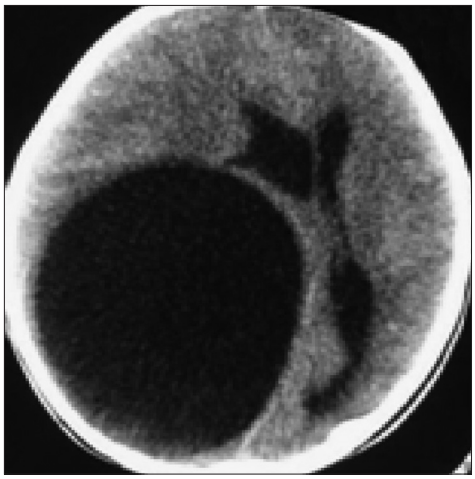

Figure 1: Solitary cystic lesion located in the right temporoparietal region

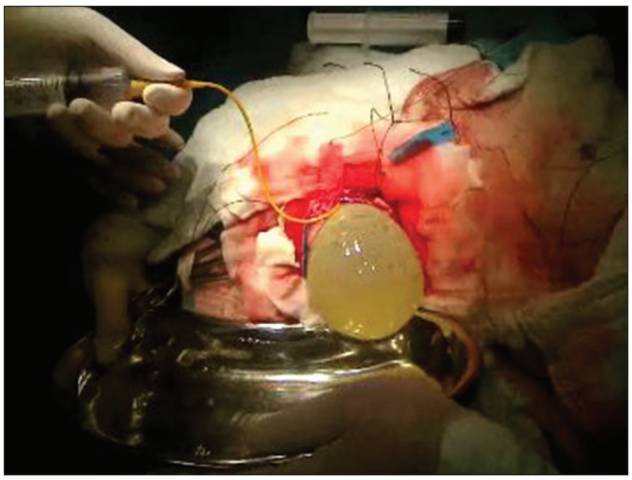

Figure 2: Appearance of the hydatid cyst during removal by the Dowling method modified by Arana-Iniguez. As the saline injection dissected the cyst from the surrounding brain, the cyst mass moved outside of the brain from its nest

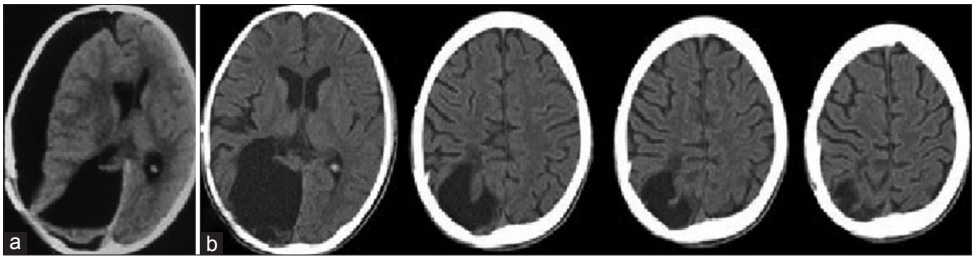

Figure 3: (a) Cranial CT showing the subdural hygroma. (b) Postoperative CT determined regression of subdural hygroma with flask, relaxed space, filled with cerebrospinal fluid

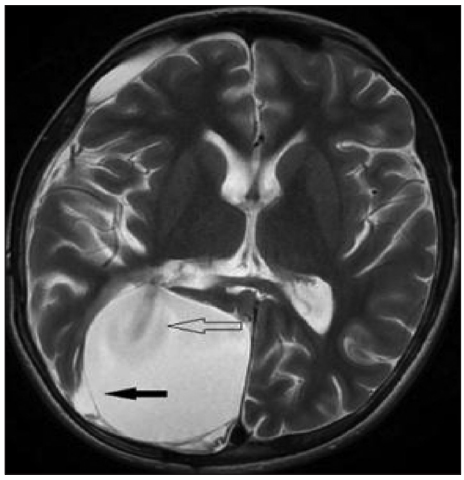

Figure 4: During follow-up, MRI scans showed a porencephalic cyst at the site where the hydatid cyst was extirpated. Interestingly, we noticed a thin outer membrane (black arrow) at the operation site around the porencephalic cavity. The one-way valve mechanism continued to fill the cyst with cerebrospinal fluid. The jet-flow (white arrow) and flow void provided evidence of the one-way filling valve

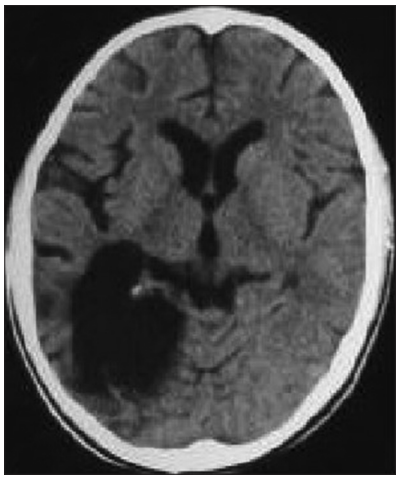

Figure 5: Postoperative CT examination showed neither a porencephalic cyst nor the surrounding membrane. The sulci around the porencephalic cyst were calm, and the operation cavity showed no tension or pressure to the surrounding tissue, thus providing evidence of decompression

cysts usually grow slowly and present clinical symptoms when they increase in size and produce a mass effect. This may be due to the elastic structure of the cranial bones,[16,17] open sutures, and compressibility of the neural tissue.[16]

The growth rate of hydatid cysts in the brain has been variably reported at 1.5-10 cm per year.[9-14] The following growth rates have been reported: Evliyaoglu *et al*.[9] reported a growth rate of 1 cm/month by assessing repeated examinations; Kemaloglu *et al*.[8] reported hydatid cyst growth rate as 4.5 cm in 6 months based on CT findings; Pasaoglu *et al*.[11] reported the growth rate of an untreated cyst as 10 cm/year based on follow-up CT; Kalelioglu *et al*.[10] reported a hydatid cyst growth rate of 5 cm/year based on follow-up CT scans; Vaquera *et al*.[14] reported a growth rate of 1 cm/year; and Sierra *et al*.[12] reported a growth rate of 5 cm/year.

Patients with intracranial hydatid cysts usually present with features such as raised intracranial pressure due to the mass effect of the large-sized cyst. Focal neurologic deficits, such as hemiparesis, visual disturbances, ataxia, seizures, or other manifestations depending on the cyst location have been reported. Headache and vomiting are almost universally present along with bilateral papilledema.[18] In our patient, headache, nausea, and vomiting were clinical symptoms and bilateral papilledema without neurologic deficits was present.

The preoperative diagnosis is very important for surgical planning and a good prognosis.[13,15,19] A study using cranial CT showed fluid that was generally the same as the CSF without perilesional edema[1]; however, Yis *et al*. reported perilesional edema,[17] which was also observed by Karak *et al*., around the cyst in the presence of active inflammation.[20] A differential diagnosis of cerebral hydatid cysts on CT, as with arachnoid cyst, porencephalic cyst, neurocysticercosis, abscess, and pilocytic astrocytoma, has been reported. Calcification of the wall is present in approximately 1% of the cysts.[18] Magnetic resonance imaging (MRI) typically shows a cystic lesion with a hypointense halo around the cyst capsule. The cyst is spherical, well defined, with thin regular margins.[19] Ozkan *et al*. reported the usefulness of CT scans in the diagnosis of cerebral hydatid disease and in the planning of appropriate surgical management.[2] We could not perform a cranial MRI prior to the operation due to the poor condition of the patient. The patient underwent an urgent operation and CT showed hypodense cystic lesion that was sharply demarcated and without perifocal edema and fluid identical to CSF. In emergencies, such as herniation in the presence of cystic lesions identified by CT, the possibility of hydatid cysts should be considered. Otherwise anaphylactic shock, chemical meningitis, or recurrence may occur due to the rupture of the hydatid cyst.[1,7,15,19,21] In literature review, we found two cases that received an urgent operation for a cerebral hydatid cyst. While Onal *et al*. treated successfully a 13-year-old girl by puncturing the cyst,[22] another patient died after operation.[23] Our patient, who also received emergency surgery due to the rapid deterioration of his neurological status, was discharged after three operations following normal neurological exams.

Intracranial hydatid cysts are usually solitary. However, 24 and 25 hydatid cysts have been reported in two different cases.[2,24] Intracranial hydatid cysts are classified as primary and secondary. The primary cysts are formed as a result of direct infestation of the larvae in the brain without the involvement of other organs. The secondary multiple cysts result spontaneously or from trauma or rupture during operation of the primary intracranial hydatid cyst.[7] Our patient's thorax and abdominal CT showed no other foci for the hydatid disease.

The main treatment for hydatid disease is complete removal of the cyst without rupture. The most commonly used surgical technique is the Dowling method modified by Arana-Iniguez.

In addition, direct puncture and aspiration of the cyst fluid through a small hole in the cyst wall or expulsion of the cyst through a small cortical incision over the cyst using insufflation of air during contralateral ventricle techniques can be applied. A small number of reports showed complete disappearance of multiple intracranial hydatid cysts with albendazole therapy using a daily dose of 10 mg/kg taken three times a day for 4 months. In the literature, recurrence rates of 19% and a mortality of 10-12%, morbidity of 9.8%, and a preoperative mortality of 8.48% have been reported. When large cysts are removed in the pediatric population, subdural collections and porencephalic cysts have been reported.[2,21] Our patient received three operations for the treatment of hydatid cysts, subdural hygroma, and fenestration of the porencephalic cyst to subarachnoid space; subsequently, postoperative albendazole therapy was applied for 4 months (10 mg/kg per day).

Conclusion

1. Hydatid cysts are known to enlarge slowly, rarely requiring emergency surgery.
2. In case of emergencies, such as midbrain herniation and if cystic lesions identified by CT, the possibility of hydatid cysts should be considered where the infection is endemic.
3. Irrespective of the preoperative diagnosis, if there is apprehension in the concrete diagnosis and suspicion of a possibility of a hydatid cyst through CT, we highly recommend the team to be prepared to remove a hydatid cyst without rupture.

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