

**CASE REPORT**

Multiple Intracranial Hydatid Cysts: A Rare Presentation

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Abstract

Cerebral hydatid disease is rare accounting for only 1-2% of intracranial space occupying lesion and usually manifests as solitary cystic nonenhancing lesions usually in the MCA territory distribution. It is even rarer to find multiple intracranial hydatid cysts or polycystic disease. We hereby report an interesting and unusual presentation of a patient with multiple intracranial hydatid cysts with a few showing inflammatory edema and minimal rim enhancement, a rare manifestation and review the literature.

Key Words

Hydatid Cysts, Echinococcus, Magnetic Resonance Imaging

Introduction

Human echinococcosis or hydatid disease is a zoonotic disease caused by tapeworms of the genus *Echinococcus*. The 2 most important forms of the disease in humans are cystic echinococcosis (hydatidosis) and alveolar echinococcosis. Humans are infected through ingestion of parasite eggs in contaminated food, water or soil, or through direct contact with animal hosts. A number of herbivorous and omnivorous animals act as intermediate hosts of *Echinococcus*. Carnivores are definitive hosts for the parasite, and are infected through the consumption of viscera of intermediate hosts that harbour the parasite and also through scavenging infected carcasses. Humans are accidental intermediate hosts and are not able to transmit the disease. Cystic echinococcosis is principally maintained in a dog-sheep-dog cycle, yet several other domestic animals may be involved including goats, swine, horses, cattle, camels and yaks. Cystic echinococcosis is globally distributed and found in every continent except Antarctica. Alveolar echinococcosis is confined to the northern hemisphere, in particular to regions of China, the Russian Federation and countries in continental Europe and North America.(1,2) In endemic regions, human incidence rates for cystic

echinococcosis can reach greater than 50 per 100 000 person-years, and prevalence levels as high as 5-10% may occur in parts of Argentina, Peru, east Africa, central Asia, and China(1,2). In India, the hydatid disease is more commonly seen in the Kurnool district of Andhra Pradesh, Madurai district of Tamil Nadu and in Punjab (3). Incidence of intracranial hydatid in India is 0.2 % (3).

Human infection with *E. granulosus* leads to the development of one or more hydatids located mainly in the liver and lungs, and less frequently in the bones, kidneys, spleen, muscles, central nervous system, and eyes. The asymptomatic incubation period of the disease can last many years until hydatid cysts grow to an extent that triggers clinical signs. Non-specific signs include anorexia, weight loss and weakness. Other signs depend on the location of the hydatid(s) and the pressure exerted on the surrounding tissues. Various series of intracranial hydatids from India have reported its incidence as 0.2% of all intracranial space occupying lesions (3, 4, 5). The typical intracranial hydatid cysts, caused by *Echinococcus granulosus*, present as well defined solitary cystic lesions in the middle cerebral artery territory (6), without surrounding edema, are non-enhancing and show

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Fig 1-3. T2 Axial Brain Images Demonstrating Multiple Cystic Lesions Which are Homogenously Hyperintense on T2 with a thin Hypointense Rim and Significant Surrounding Edema and Moderate Mass Effect; Axial FLAIR Image of the Same Patient Demonstrating Multiple Homogenous Hypointense Clesions with Significant Edema.; Gadolinium Enhanced Axial Brain MRI Images Demonstrating Minimal peripheral rim enhancement of the above mentioned lesions

unremarkable mass effect unless large (7) when they can show significant parenchymal distortion, mass effect, hydrocephalus, and raised intracranial pressure. Lesions can present as multicystic masses, with mass effect or with edema, when differentiation may be difficult from other entities like astrocytoma, infective lesions etc. (7). Multiple cysts are extremely rare and can develop either spontaneously, after trauma or post operatively. We present a case with multiple hydatid cysts in bilateral cerebral hemispheres, a rare manifestation.

Case Report

A 35 years gentleman, a businessman by profession, presented to us with complaints of progressive weakness of right upper limb since 2 weeks. No history of any trauma, falls, loss of consciousness or any positive motor phenomenon was reported. No history suggestive of headaches, visual obscuration, vomiting was elicited. Patient did not report any constitutional symptoms like cough, fever, significant weight loss or any history of promiscuous behavior. History of excision of an intracranial cyst in the right occipital lobe was reported 7 months back which was diagnosed to be hydatid cyst after biopsy and other serological investigations. No history of any complications during surgery was reported and post-operative period had remained uneventful. Patient had been further managed with albendazole for the next 6 months which had been discontinued only a month back. Examination revealed a conscious and alert patient with normal vitals. Pupils were 3 mm in size bilaterally with normal reaction to light and fundus did not reveal any signs of papilloedema. Extraocular movements were

full. Right supranuclear facial palsy was observed and rest of the cranial nerves were normal. Motor examination revealed grade 2 power in right upper limb and grade 4 power in right lower limb and right plantar was extensor. NCCT head revealed multiple large, oval, hypointense cystic lesions of CSF density in bilateral cerebral hemispheres without significant mass effect or midline shift. MRI brain revealed multiple cystic lesions. The cyst contents were slightly hyperintense to CSF on T1WI and hyperintense on T2WI (Fig 1). A couple of lesions showed moderate perilesional edema on FLAIR sequence (Fig 2) with minimal enhancement on contrast enhancement with gadolinium (Fig 3). A lesion in the left frontoparietal region especially revealed significant perilesional edema and mass effect accounting for the right arm and leg weakness. A possibility of multiple intracranial hydatidosis was considered which may be primary or possibly secondary to seeding during the previous surgery. CT chest and abdomen was performed which revealed multiple cystic lesions in the liver and further management with excision of the left parietal cyst along with medical therapy was considered.

Discussion

Intracranial hydatid disease is rare, with reported incidence of 1-2% of all cases with hydatid disease (1). The parietal lobe is the commonest site. All four cases reported by Dharker *et al* (3) and three out of five cases of intracerebral hydatid cysts reported by Balasubramaniam *et al* (8) had parietal lobe involvement. The other less common sites reported are skull, cavernous sinus, eye ball, pons, skull, extradural, cerebellum and

ventricles (9). Intracranial hydatid cysts are commonly solitary. Multiple intracranial cysts are rare. Onal et al found only three cases of multiple cysts in their series of 33 cases (2). Intracranial hydatid cyst 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. 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. Primary multiple cysts are uncommon and isolated case reports of primary multiple hydatid cysts have appeared in the literature. Nurchi et al (10) while reviewing the literature found only eleven reported cases of primary multiple hydatid cysts. The cerebral hydatid cysts are slow growing and present late when they increase in size and become large. There is no consensus on the growth rate of the hydatid cysts of the brain and has been variably reported between 1.5-10 cm/year (3). Formation of the large multiple hydatid cysts within seven months of the first surgery following total excision in our patient suggests that probably the growth rate is higher. Features of raised intracranial pressure is the commonest manifestation which may be due to the large size or due to interference with pathway of CSF flow. Seizures is the second commonest manifestation. Erashin et al (1) observed that 18 out of 19 cases presented with raised intracranial pressure. Four cases had seizures. Our patient had only hemiparesis without any features of raised intracranial pressure. MR and CT scans characteristically show hydatid cyst as a spherical, well defined, non enhancing cystic lesion without peripheral oedema (11) The fluid density is generally equal to that of CSF on both CT and MR scan. A fine rim of peripheral enhancement with perilesional oedema may be seen in the presence of active inflammation. MR scan may show a low density cyst wall (11) and relations with surrounding structures is better delineated than on CT scan. The treatment of hydatid cyst is surgical and the aim of surgery is to excise the cyst in toto without rupture to prevent recurrence and anaphylactic reaction (12). 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 insufflation of air in the contra lateral ventricle and the most commonly done procedure designed to give birth to the intact cyst by irrigating saline between cyst wall-brain interface. This is possible because of minimal adhesions around the cyst wall. Only a few reports are available mentioning the efficacy of drug therapy. Isolated case reports showed complete disappearance of multiple intracranial hydatid cysts with Albendazole therapy in a daily dose of 10 mg/kg, taken three times a day for four months. Erashin et al (1) reported better effectiveness of the drug therapy in recurrent cases and in cases with rupture at the surgery.

Conclusion

Cerebral hydatid disease should be suspected especially when cystic lesions appear in the CNS. The diagnosis should be suggested by evidence of a primary hepatic focus, appropriate clinical history, high prevalence of the infection in the host's geographic location, and laboratory findings. Serologic analysis is in most cases negative; pathohistologic analysis is the most reliable method of diagnosis.

References

1. Erashin Y, Mutluer S, Guzelbag E : Intracranial hydatid cysts in children. *Neurosurgery* 1993; 33 : 219-224.
2. Onal C, Orhan B, Metis O, et al . Three unusual cases of intracranial hydatid cysts in paediatric age group. *Pediatr Neurosurg* 1997; 26 : 208-213.
3. Dharker SR : Hydatid disease. In : Text Book of Neurosurgery, 2nd edition. Eds. Ramamurthi B, Tandon PN. Churchill Livingstone, New Delhi 1996. pp. 535-544.
4. Raja Reddy D, Dayanand Rao B, Prabhakar V et al : Hydatid disease of the central nervous system. *Indian J Surgery* 1972; 34 : 192.
5. Sardana VR, Dharker SR, Mittal RS, et al . Multiple intracranial hydatid cysts. *Neurol India* 1991; 39 : 205.
6. Ba'assiri A, Haddad FS. Primary extradural intracranial hydatid disease. CT appearance. *AJNR* 1984; 5: 474 -475.
7. Pinar P, Mecit K, Fatih A, et al. Hydatid disease from head to toe. *Radiographics* 2003;23:475-494.
8. Ahmet M, Erhan Türkolu, MD, Hayri K, Bora G. Hydatid cyst of the ambient cistern radiologically mimicking an arachnoid cyst. *Journal of Neurosurgery Pediatrics* 2012; 10:3, 186-88. ..
9. Jimenez-Mejias ME, Castillo-Ojeda E, Cuello-Contreras JA, et al. Cerebral hydatidosis. Analysis of a series of 23 cases. *Med Clin (barc)M* 1991; 97 : 125-32.
10. Nurchi G, Francesco F, Montaldo C et al : Multiple cerebral hydatid disease : case report with magnetic resonance imaging study. *Neurosurgery* 1992; 30 : 436-38. ..
11. Coates R, Von Sinner W, Rahm B. MR imaging of an intracranial hydatid cyst. *AJNR* 1990; 11 : 1249-50.
12. Duishanbai S1, Geng D, Liu C, et al. Treatment of intracranial hydatid cysts. *Chin Med J (Engl)* 2011 ;124(18):2954-8.