Camalote sign in intraventricular hydatid cyst: A rare presentation of uncommon disease

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INTRODUCTION

Cerebral hydatid cysts involve <2% of intracranial space-occupying lesions, usually located in supratentorial white matter.[5] It was first described in 1550 by Paranoli in the corpus callosum.[6] Other rare sites described include ventricles, cisterns, brainstem, basal ganglion, and extradural sites. Literature review suggests that less than 40 cases are described for intraventricular location of hydatid cyst but only one case report has revealed floating water lily sign (also called Camalote sign) in an intraventricular cyst.[⁴,⁹,¹⁰] Here's description of one such rare presentation of intraventricular hydatid cyst with evident Camalote sign in radiological image. It's appropriate management including surgical approach and resection is also elaborated.

CASE REPORT

A 5-year-old male child was brought to our department with chief complaints of holocranial headache, irritability, occasional vomiting for 3 months, and altered sensorium for 3 days. On examination, patient was in altered sensorium with GCS of E3V4M6. Magnetic resonancace
imaging (MRI) brain were suggestive of a large ruptured intraventricular cyst with irregular floating cyst wall in the right frontal horn giving a typical Camalote sign appearance [Figure 1]. It was causing obstruction of foramen of Monro with gross hydrocephalus. On the basis of MRI findings working diagnosis of ruptured hydatid cyst was made. Under steroid cover, Patient was planned for transcortical approach as the lesion was present in the frontal horn of the right lateral ventricle causing its ballooning and thinning out of the overlying cortex. Patient underwent right frontal craniotomy and ventricle was opened up through transcortical approach to access the cyst, taking special care to prevent spillage of cyst contents. The cyst wall was free-floating in clear CSF. Cyst was deroofed and cyst contents were aspirated followed by instillation of hypertonic saline in the cyst wall and aspiration again. After completely emptying the cyst of its contents, its wall was dissected out [Figure 2]. Ultrasonogram of the abdomen and X-ray of the chest were done to evaluate for any other primary lesion, but these didn’t reveal any pathology.

Histopathology also confirmed the diagnosis [Figure 3]. Patient recovered well after the surgery and there was no need of CSF diversion. He was given three cycles of albendazole therapy of 1 month duration after intervals of 14 days between each cycle. Patient symptoms also regressed and there was no recurrence of the cyst during 3 years of follow-up.

**DISCUSSION**

Hydatidosis is common endemic zoonotic condition in grazing areas of world caused by flatworm cystodes: *Echinococcus*

**Figure 1:** (a-c) T1 MRI axial image sequences showing hypointense intraventricular cyst 1 (d-f) T2 MRI axial image sequences showing hyperintense intraventricular cyst with ruptured hypointense wall in the right lateral ventricle (Camalote sign) 1 (g-i) T2 MRI Saggital image sequences again showing the ruptured cyst and cyst wall sedimenting down 1 (j-l) T2 MRI Coronal image sequences confirming the ruptured hydatid cyst. (m and n) MRI FLAIR images in axial cuts distinctly defining the cyst wall.

**Figure 2:** Excised ruptured hydatid cyst wall.

**Figure 3:** (a) Lamellated membranes of hydatid cyst with scolex (blue arrow) (H and E, ×100) 3 (b) High power photomicrograph of cross-section of a scolex at the level of invaginated head bearing the hooklets (red arrow).
granulosus (unilocular) and Echinococcus multilocularis (alveolar form). Humans as accidental hosts, ingest larval form of Echinococcus with contaminated food. From the intestine, they are taken to liver through portal circulation, from where they may spread to different parts of body through systemic circulation. Common site of involvement is intraparenchymal especially MCA territory distribution (most common in parietal lobes) where it reaches as embolic spread. Case reports including other uncommon sites (ventricles, cisterns, skull, brainstem, cerebellum, orbits, and extradural sites) exist in literature. On extensive literature search, we could find two important review articles summarising the previously reported intraventricular hydatid cyst which comprise total of 30 cases in large series and 12 cases as individual case reports. Echinococcus oncospheres reach ventricles by embolic spread to choroid plexus. Hydatid cysts in any part of the body have three layers. (i) Outer pericyst: layer made from fibrous tissue deposited by host due to granulomatous reaction around the cyst. (ii) Ectocyst: the acellular layer which allows passage of nutrients. (iii) Endocyst: layer comprising the thin and translucent germinal layer and the laminated membrane which produces scolecis. Endocyst secretes clear fluid into the cystic cavity. This fluid is rich in various electrolytes, proteins, lipids, and polysaccharides and is highly antigenic which may cause anaphylactic reaction if it comes in contact with normal host tissues. World Health Organisation has classified pericyst being made up of host tissues without surrounding edema. The cyst wall do not enhance with contrast imaging except in rare cases having secondary infection in cyst. The WHO classifies hydatid cyst in the six stages on basis of its evolution [Table 1].

Computed tomography (CT) and MRI features of hydatid cyst are of a spherical well defined, thin walled, homogenous cystic lesion with CSF like fluid density. These imaging features mandates ruling out differential diagnosis of arachnoid cyst, dermoid or epidermoid cyst. Calcification or septations may be present rarely. Presence of daughter cyst is pathognomonic for hydatid cyst but uncommon in radiology. Conventional water lily sign is present on ultrasound, X-rays, CT scan or MRI in stage 3A when the endocyst wall is detached from the pericyst and floats freely into the cystic fluid. This sign has been well described in hepatic and pulmonary hydatid cyst but very rare in CNS hydatidosis. Sadashiva et al. described this sign in intraventricular hydatid cyst as a floating water lily sign. It was described in postintervention MRI when the cyst got ruptured iatrogenically during CSF diversion. In other case report by Thakur et al. in 2017, there was spontaneous rupture of the cyst and multiple daughter cysts were seen in temporal and occipital horns. Although typical Camalote sign was not described however imaging in that case report appeared as classical water lily sign. In our case, there was rupture and complete separation of endocyst wall giving a typical floating water lily sign with intact outer layers. Clinical presentation of hydatid cyst is mainly due to the mass effect caused by the cyst on surrounding structures commonly leading to raised ICP.

The conventional Dowling’s technique has been described with an aim to take out the intact cyst by hydrostatic expulsion holds good for parenchymal cysts. However, intraoperative rupture has been described in around 25% of cases even with the use of this technique. Especially for large and deep-seated cysts including intraventricular one, there are more complications like intraventricular/parenchymal hemorrhage, subdural effusions, and meningitis. Few authors have described opening/puncturing the cyst and aspiration of cyst contents followed by removal of the shrunken cyst wall to avoid such complications which can be considered as modification of PAIR technique (percutaneous aspiration, injection, and respiration) which is widely accepted for hepatic hydatid cysts. Albendazole is always recommended in all such cases for several months with 1 month cycle of albendazole followed by 14 days of therapy free intervals.

**CONCLUSION**

Intraventricular location of hydatid cyst is very uncommon but it should be kept as a differential diagnosis especially in endemic areas. The presence of camalote sign on imaging in such rare sites is very helpful in confirming the diagnosis preoperatively for appropriate surgical planning in such cases.

**Declaration of patient consent**

Patient’s consent not required as patients identity is not disclosed or compromised.
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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Copley IB, Fripp PJ, Erasmus AM, Otto DD. Unusual presentations of cerebral hydatid disease in children. Br J Neurosurg 1992;6:203-10.
2. Golzari SE, Sokouti M. Pericyst: The outermost layer of hydatid cyst. World J Gastroenterol 2014;20:1377-8.
3. Grecu I, Rizea O, Lupescu IG. The Water Lily Sign: A Specific and Valuable Imaging Feature Poster No. C-1246, Congress: ECR 2017 Infection; 2017.
4. Guzel A, Tatli M, Maciaczyk J, Altinors N. Primary cerebral intraventricular hydatid cyst: A case report and review of the literature. J Child Neurol 2008;23:585-8.
5. Hajhouji F, Aniba K, Laghmari M, Lmejjati M, Ghannane H, Benali SA. Epilepsy: Unusual presentation of cerebral hydatid disease in Children. Pan Afr Med J 2016;25:58.
6. Krüger J, Ritz A, Ingunza W. A case of intraventricular hydatid cyst. In: Frowein RA, Wilcke O, Karimi-Nejad A, Brock M, Klinger M, editors. Head Injuries. Tumors of the Cerebellar Region. Advances in Neurosurgery, Vol. 5. Berlin, Heidelberg: Springer; 1978.
7. Oscar H, Brutto D, Figueroa JE, Garcia HH. Parasaitic infections. In: Youmans and Winn Neurological Surgery. 7th ed. Amsterdam, Netherlands: Elsevier; 2017. p. 354-5.
8. PAIR: Puncture, Aspiration, Injection, Re-Aspiration. An Option for the Treatment of Cystic Echinococcosis; 2021.
9. Pandey S, Pandey D, Shende N, Sahu A, Sharma V. Cerebral intraventricular echinococcosis in an adult. Surg Neurol Int 2015;6:138.
10. Sadashiva N, Shukla D, Devi BI. Rupture of intraventricular hydatid cyst: Camalote sign. World Neurosurg 2018;110:115-6.
11. Singounas EG, Leventis AS, Sakas DM, Lampadarios DA, Karvounis PC. Successful treatment of intracerebral hydatid cysts with albendazole: Case report and review of the literature. Neurosurgery 1992;31:571-4.
12. Stojkovic M, Rosenberger K, Kauczor HU, Junghanss T, Hosch W. Diagnosing and staging of cystic echinococcosis: How do CT and MRI perform in comparison to ultrasound? PLoS Negl Trop Dis 2012;6:e1880.
13. Thakur SH, Joshi PC, Kelkar AB, Seth N. Unusual presentation of hydatid cyst-ruptured intraventricular hydatid. Indian J Radiol Imaging 2017;27:282-5.

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